Primary Intraosseous Cavernous Hemangioma in the Skull

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Abstract: Primary intraosseous cavernous hemangiomas (PICHs) are benign vascular tumors that may involve any part of the body. PICH occurs more frequently in the spine and less commonly in skull. The earliest description in the English literature was in 1845 by Toynbee, who reported a vascular tumor arising in the confines of the parietal bone. Skull PICHs do not always have typical radiologic features and should always be considered in the differential diagnosis of malignant skull lesions. We now reviewed and analyzed related literatures in detail with reporting a rare case of PICH in the left front bone that was surgically resected.

INTRODUCTION

Primary intraosseous cavernous hemangiomas (PICHs) is a rare bone tumor accounting for 0.7% to 1.0% of all bone tumors.1 PICHs are usually found in the vertebral column and rarely seen in the skulls. The earliest description of skull PICH was in 1845 by Toynbee. Much to our knowledge, there have only been 93 cases of skull PICH reported previously. The number of relevant literatures each year shows a general tendency to increase over time. A timeline of the related publications is available as Figure 1. On the basis of a world map with the global distribution of skull PICH-related publications based on the analysis of their geolocational data, the countries that the publications are from are mainly concentrated in Europe, North America, and East Asia (Figure 2).

PICH is mostly seen in middle age and the male-to-female ratio ranges from 3:1 to 2:1.2,3 Among the skulls, the frontal bone is the most commonly involved, followed by the parietal bone, temporal bone, and less frequently by the occipital bone. The pathogenesis is still unknown but a history of trauma seems to be related in some case reports.4 Total surgical excision is the treatment of choice and the prognosis after complete excision is excellent and recurrence is usually rare. Herein, we present a rare case of a skull PICH in a 17-year-old girl. The clinical presentation, pathology, differential diagnosis, and treatment of this rare disorder are discussed. Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the editor of this journal. Because of this, there is no need to conduct special ethic review and the ethical approval is not necessary.

CLINICAL CASE DESCRIPTION

A 17-year-old girl presented with a swelling on the left forehead, which had been slowly enlarging for 2 year. She denied headache, dizziness, and past trauma history. Other medical history and a review of systems were insignificant. On examination, the diameter of the mass on the left frontal bone was about 6 cm long. In consistency, the lesion was immobile, hard, and not tender. There is no abnormality for routine biochemical tests.

Plain cranioural x-ray demonstrated a radiolucent mass and the CT image showed a 9.3 mm × 18.9 mm × 13.4 mm osteolytic lesion within the diploe on the frontal bone near the orbital roof (Figure 3). Original impression of the mass by a radiologist was a skull eosinophilic granuloma or a fibrous dysplasia. A left parietal craniectomy and total lesion resection with a margin of surrounding normal bones was conducted under general anesthesia. The lesion extended intracranially to the adjacent dura mater loosely and externally to the corresponding scalp. It was rich in blood supply by clusters of small vessels. After surgery, a histologic examination of the specimen demonstrated an intradiploic cavernous hemangioma featured by expanded small blood vessels with thin wall and sinusoids surrounded by a thin layer of endotheliocytes. The patient had a good recovery after surgery. At 1-year follow-up there was not any lesion recurrence.

DISCUSSION AND REVIEW

Hemangiomas can be histologically divided into 3 types: cavernous, capillary, and mixed. Cavernous hemangioma consists of clusters of dilated blood vessels, which are separated by fibrous septa, whereas capillary ones are rich in small vascular luminas without much fibrous septa. The majority of hemangiomas in skull are of the cavernous type (PICHs), while hemangiomas in vertebrae are usually the capillary type. PICHs of the cranium are rare benign vascular tumors that account for about 0.2% of all bone tumors and 10% of benign skull tumors.1 It occurs most commonly in the vertebral column and rarely in the skull. Of the 93 cases of skull PICH reported in previous literatures from 1845 to 2015, 44.1% were located in the frontal bone, 12.9% involved the temporal bones, 11.8% occurred in the occipital bone, 12.9% in parietal bone, and 5.4% in Cranial fossa; fewer cases have been reported in sphenoid, zygomatic, ethmoid, clivus, and orbital rim, etc. (Table 1 and Figure 4).

They are predominantly seen in patients in their fourth and fifth decades. Unlike the age predominance, our patient was an adolescent female (17-year old). The male-to-female ratio...
FIGURE 1. A timeline of the publications related to skull PICH.

FIGURE 2. A world map with the global distribution of skull PICH-related publications based on the analysis of their geolocational data.

FIGURE 3. CT scan (bone window) demonstrated an intradiploic osteolytic mass.
ranges from 3:1 to 2:1.² The earliest description in the English literature was in 1845 by Toynbee, who reported a vascular tumor arising in the confines of the parietal bone.⁴ PICH arises from the vessels in the diploic space and supplied by the branches of the external carotid artery, arising in the skull vault. The middle and superficial temporal arteries are the main sources of blood supply. Within the lesion, the capillaries are widely dilated separated by fibrous tissue. The pathogenesis of PICH remains unknown. The cause is considered to be congenital, but this has not yet been proven. Some scholars proposed a hereditary nature for “vascular malformations” in the skull with an autosomal recessive inheritance mode. Others indicated that proliferation and differentiation of the undifferentiated primitive mesenchymal cells induced by a variety of stimuli may be the potential etiology. Trauma may also be an important etiology.⁵

Like our present case, the patient presented with a slow-developing palpable firm swelling without tenderness. Local neurological deficits were uncommon partly because the masses

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**TABLE 1. Literature Review of Cases of Intraosseous Hemangioma of the Skull From 1845 to 2015 (Total 93 cases)**

| Frontal | Temporal | Occipital | Parietal | Cranial Fossa | Other Sites |
|---------|----------|-----------|----------|---------------|-------------|
| Pilcher, 1894 | Sharma et al, 1999 | Sargent et al, 1965 | Petersen et al, 1992 | Toynbee et al, 1845 | Jackson et al, 1980 |
| Wyke, 1949 | Suzuki et al, 2001 | Mangham et al, 1981 | Cervoni et al, 1993 | Kuman et al, 1993 | Glasscock et al, 1984 |
| Gupta et al, 1975 | Heckl et al, 2002 | Mangham et al, 1981 | Corr, 2000 | Yoshida et al, 1999 | Mazzoni et al, 1988 |
| McIntyre et al, 1977 | Pottelbergh et al, 2004 | Mangham et al, 1981 | Garci?a-Mar et al, 2001 | Heckl et al, 2002 | Bottrill and Poc, 1995 |
| Gross and Roth, 1978 | Politi et al, 2005 | Glasscock et al, 1984 | Khanam et al, 2001 | Paradowski et al, 2007 | Khaman et al, 2001 |
| Fouad et al, 1979, ×2 | Cheng et al, 2006 | Suss et al, 1984 | Heckl et al, 2002 | Naama et al, 2008 | Naama et al, 2008 |
| Shinno et al, 1986 | Buhl et al, 2007 | Mazzoni et al, 1988 | Buhl et al, 2007 | Sasagawa et al, 2009 | Sasagawa et al, 2009 |
| Hook et al, 1987 | Nasser et al, 2007 | Buchanan et al, 1992 | Mazzoni et al, 1988 | Gibson, 2007 | Ruma et al, 2013 |
| Zucker et al, 1989 | Naama et al, 2008, ×2 | Fierek et al, 2004 | Prayson, 2007 | Atci et al, 2013 | Atci et al, 2013 |
| Hoffmann et al, 1990 | Sasagawa et al, 2009 | Fierek et al, 2004 | Sasagawa et al, 2009 | Prakash et al, 2011 | Lee et al, 2014 |
| Hornig et al, 1990, ×2 | Roel et al, 2012 | Yang et al, 2014 | Balzara et al, 2008 | Hsiao et al, 2015 | Hsiao et al, 2015 |
| Sinnreich, 1990 | Park et al, 2013 | Yeti et al, 2014 | Hsiao et al, 2015 | Kilani et al, 2015 | Kilani et al, 2015 |
| Aurora et al, 1991 | Xu et al, 2013 | Uemura et al, 2014, ×2 | Murrone et al, 2014 | Chun et al, 2015 | Yen et al, 2014 |
| Faerber and Hiatt, 1991 | Relf et al, 1991, ×2 | Peterson et al, 1992 | Cervoni et al, 1995, ×2 | Pastore et al, 1999 | Pastore et al, 1999 |
| 41 cases (44.1%) | 12 cases (12.9%) | 11 (11.8%) | 12 (12.9%) | 5 (5.4%) | 12 (12.9%) |

Other sites indicate the case where the lesion was located in sphenoid, zygomatic, ethmoid, clivus, and orbital rim, etc.

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**FIGURE 4.** Pie graph of the location of previous reported skull intraosseous hemangioma cases.
| Author     | Year | Age, y | Sex | Clinical Features                                                   | Size                | Location                  | Radiographic Features | Treatments                      | Pathologic Features                                                   | Outcomes                                                                 |
|------------|------|--------|-----|--------------------------------------------------------------------|---------------------|---------------------------|-----------------------|---------------------------------|------------------------------------------------------------------------|--------------------------------------------------------------------------|
| Muzumdar   | 2001 | 26     | F   | A bony swelling over the occipital region in the region of the torcular area, progressively enlarging for 5 years | 8 cm x 8 cm x 6 cm  | Occipital bone            | CT showed an intradiploic expansive tumor in the occipital region in close proximity to the torcular area, with a characteristic sunburst pattern with striations radiating from the center | Partial resection and radiotherapy                                      | Multiple dilated vascular spaces lined by endothelium separated by fibrocollagenous tissue | The patient was well at follow-up examination after 9 months |
| Muzumdar   | 2001 | 30     | F   | A painful occipital swelling, progressively enlarging for 8 months. | 7 cm x 3 cm x 3 cm  | Occipital bone            | Axial CT showed an intradiploic subtorcular tumor expanding the inner and outer tables | Preoperative embolization and partial excision                        | Histological examination confirmed the diagnosis of cavernous hemangioma | The patient was well and symptom-free at follow-up examination after 12 months |
| Friedman   | 2002 | 51     | M   | Recurrent episodes of right facial spasm for 3 years and recurrent right facial paresis for 2 years | –                   | Right temporal bone       | CT revealed a soft tissue mass of the right temporal bone, involving the geniculate ganglion, with extension to the tympanic segment of the facial nerve | Observation for 2.5 years. Then, he underwent excision of the mass through a right transmastoid and middle fossa approach. The facial nerve was resected along with the mass. A greater auricular nerve cable graft was used to repair the facial nerve. | Histopathologic evaluation showed that the lesion was a cavernous hemangioma of the facial nerve | At 12 months after surgery, his right-sided facial function had improved |
| Liu        | 2003 | 40     | F   | Chronic headaches and facial numbness                              | 2.5 cm              | Left sphenoid bone at the skull base                             | MRI showed a 2.5 cm enhancing mass involving the left sphenoid bone at the skull base, encroaching on the cavernous sinus and mildly displacing the cavernous carotid artery posteriorly | The patient elected surgical removal for a definitive diagnosis and complete resection | Thin-walled vascular channels lined by a single layer of flattened endothelial cells interspersed among bony trabecula | Her facial numbness resolved in the ensuing weeks and her headache symptoms improved dramatically. The patient has remained stable after 1 year of follow-up |
| Dogan      | 2005 | 53     | F   | Stabbing headache in the right frontal region and local swelling  | 6 cm x 6 cm x 3 cm  | Right frontal bone        | CT demonstrated an extensive lesion that had eroded both tables of the right frontal bone. The mass had well-defined margins, and a mixed lytic, and slightly sclerotic appearance | Surgery was performed with en bloc resection of the lesion and additional removal of a 1-cm-wide margin of the surrounding uninvolved bone | Multiple endothelium-lined vessels of varying sizes embedded in fibrolipomatous tissue | The patient did well after the operation. Follow-up at 6 months post-surgery revealed no recurrence |
| Author     | Year | Age, y | Sex | Clinical Features                                                                 | Size             | Location                  | Radiographic Features                                                                 | Treatments                          | Pathologic Features                                                                 | Outcomes |
|------------|------|--------|-----|----------------------------------------------------------------------------------|------------------|---------------------------|---------------------------------------------------------------------------------------|-------------------------------------|-------------------------------------------------------------------------------------|----------|
| Politi     | 2005 | 46     | F   | A gradually enlarging mass on her forehead. The mass was painless and did not produce any symptoms except cosmetic deformity | 3 cm × 4 cm × 6 cm | Right frontal bone        | MRI demonstrated a mass in the right frontal bone with intra- and extracranial extension with an enhanced dural tail after gadopentetate dimeglumine injection | Complete resection                   | Dilated blood-filled vessels, arranged in a diffuse haphazard pattern, with a single layer of endothelial cells | —        |
| Nasser     | 2007 | 32     | M   | A swelling above the right eyebrow persisting for 1 year                          | 1.5 cm           | Lateral to the right frontal sinus | CT demonstrated a 1.5-cm mass between the outer and inner tables just lateral to the right frontal sinus. The outer and inner tables were thin and partially defective | The lesion was removed en bloc, and the circumferential normal part of the surrounding bone was also removed | Photomicrograph showing thin-walled vascular channels lined by a thin layer of endothelial cells interspersed among bony trabeculae | —        |
| Paradowski | 2007 | 35     | F   | Headaches for 6 years                                                            | –                | Left parietal bone         | X-ray showed an osteolytic lesion in the parietal bone, suggesting the presence of hemangioma. CT and MRI: honeycomb-like internal structure, pressing the adjacent cortex | Neurosurgery of the tumor involved total resection and cranioplasty | Immunohistochemical examination revealed the presence of endothelial cells expressing CD34 and vimentin | The brain compression resolved and the headaches improved |
| Reis       | 2008 | 73     | M   | A sudden onset of dizziness and reading difficulty                               | –                | Right frontotemporal region | An ischemic area on the left occipital region and an intraosseous expansive lytic lesion on the right frontotemporal region, without signs of brain tissue involvement | Right frontotemporal craniectomy with en bloc resection of the osseous lesion, followed by cranioplasty with acrylic cement | Primary osseous cavernous-type hemangioma | –        |
| Cosar      | 2008 | 16     | M   | A complaint of painless swelling lesion on the right part of his calvarium skull that had developed in the previous 3 months | 2 cm × 2 cm × 2 cm | Right parietal bone        | CT revealed an intradiploic mass in the right parietal bone. T1WI showed nonhomogeneously isointense mass and contrast enhancing while T2WI revealed a heterogeneous hyperintense images | The mass was removed with a rim of surrounding normal bone tissue | The photograph showing the expanded vascular structures layered with endothelial cells between the bone spicules | Postoperative course of the patient was uneventful. Three months follow-up CT and MRI of the patient revealed no recurrences and complaints |
| Author          | Year | Age, y | Sex | Clinical Features                                                                 | Size | Location                  | Radiographic Features                               | Treatments                                                                 | Pathologic Features                                                                 | Outcomes                                                                 |
|-----------------|------|--------|-----|-----------------------------------------------------------------------------------|------|---------------------------|-----------------------------------------------------|---------------------------------------------------------------------------|--------------------------------------------------------------------------------|--------------------------------------------------------------------------|
| Sasagawa        | 2009 | 55     | F   | Presented with a painless swelling on her right forehead 6 years previously        | 2 cm | Right forehead            | CT demonstrated an osteolytic lesion. The lesion    | The lesion was resected en bloc and the circumferential normal bone was    | Erythrocytes filling many sinusoidal channels lined with a single layer    | The postoperative period was uneventful at follow-up examination after 2   |
| Salunke         | 2010 | 11     | M   | Progressive painless bilateral visual loss over the last 2 years; chronic headache; blind in both eyes and fundoscopy showed optic atrophy | –    | Skull base                | CT revealed an abnormal rarefaction and trabeculated appearance with intact cortical margins, extending inferiorly into the pterygoid plates and anteriorly into the ethmoidal sinuses. MRI showed a huge mass, isointense on T1, hyperintense on T2 and enhancing uniformly on contrast | Bilateral internal maxillary arteries were embolized using PVA particles (300–700 µ). The tumor was debulked via an extended bifrontal approach | Thin-walled vascular channels lined by flattened endothelial cells interpersed among bony trabeculae diagnostic of intraosseous cavernous hemangioma | The patient succumbed to post operative infection                           |
| Tyagi           | 2011 | 28     | F   | Huge swelling over the right parietal region progressively increasing in size over the past 15 years | 6.3 cm \times 5.3 cm \times 5.6 cm | Right high parietal region | CT: a uniform hyperdense mass. The involved bone showed complete erosion of both the inner and outer tables | Complete resection Bone bits with mature lamellar bone and bony spicules. Medullary spaces between bony trabeculae showing ectatic thin-walled blood vessels with single layer of flat endothelial cells | Symmetric expansion of cancellous bone covered by an unremarkable cortex with normocellular intervening marrow consistent with reactive bone. Numerous irregular small blood vessels permeating the mature fibrous tissue | At 3-year follow-up there is no recurrence of lesion                       |
| Philpott        | 2011 | 1      | F   | An enlarged but stable head circumference greater than the 95th percentile with no neurologic signs | –    | Right parietal bone       | MRI: an unexpected finding of focal expansion of the cancellous diploe of the right parietal bone with diffuse thickening and enhancement of the underlying dura | Excisional biopsy, along with resection of abnormal bone, was performed | Symmetric expansion of cancellous bone covered by an unremarkable cortex with normocellular intervening marrow consistent with reactive bone. Numerous irregular small blood vessels permeating the mature fibrous tissue | MR imaging at 6-month follow-up revealed reduction in dural thickening consistent with spontaneous involution and appropriate bony healing |
| Author | Year | Age, y | Sex | Clinical Features | Size | Location | Radiographic Features | Treatments | Pathologic Features | Outcomes |
|--------|------|--------|-----|------------------|------|----------|----------------------|------------|---------------------|----------|
| Yucel  | 2011 | 4 months | F   | Neurological examination revealed no abnormality except the right parietal solid mass lesion | 3 cm | Right parietal bone | CT showed a soft tissue density mass that expanded the diploic space on the right side. Both inner and outer tables were increased in thickness and there were some defective areas on the cortex of the inner table | A right parietal craniectomy with total excision of the lesion and a margin of surrounding uninvolved bone | Dilated and proliferated vascular space between the bone trabeculae. Monoclonal antibody to CD34 highlights a single layer of flattened endothelial cells lining the dilated vascular spaces | – |
| Xu     | 2013 | 24     | M   | Progressive right eye exophthalmous for 1 year; left frontal bone destruction was found on plain radiographs of the skull | 3 cm × 3 cm × 3 cm | Left frontal bone | CT: a mass with regular edges destroying the surrounding bone and penetrating into the internal skull in the upward wall of the right orbit. MRI: T1-isointense and T2-hyperintense circumscribed lesions. These uniformly enhancing lesions were associated with an enhancing dural tail | Complete resection | Immunohistochemical staining for CD34, CD31, P53, and Ki-67 were all positive | – |
| Rumana | 2013 | 3 months | M   | Have a small soft swelling over the parietal area at birth. No history suggestive of perinatal insult. The swelling increased in size and became harder | 5 cm × 6 cm | Right parietal area | Asymmetric enlargement of the skull over the right parietal area with increased diploic space of the right parietal bone. The inner table was irregular and the outer table was thinned and expanded | Parietal craniectomy with total excision of the lesion | Thin-walled, dilated, irregular vascular channels lined by a single layer of flattened endothelial cells interspersed among bony trabeculae, consistent with an intraosseous cavernous hemangioma | – |
| Aki    | 2013 | 38     | M   | A left parietal mass and intermittent localized pain was admitted to our clinic. A history of a motor vehicle injury to the left parietal region 2 years ago | 2 cm × 2 cm × 2 cm | Left parietal bone | CT: an increase in the left parietal diploic space. MRI: an expansive lesion in the bone structure | Complete resection | Fibroconnective tissue containing osseous areas in some places and large blood vessels in the stroma | – |
| Park   | 2013 | 39     | F   | A bony hard, tender swelling mass on the left forehead, which had been slowly enlarging for 1 year. She had a history of minor trauma on her forehead 1-and-a-half years earlier | 3 cm × 3.5 cm × 1.5 cm | Left frontal bone | CT scan demonstrated an intradiploic osteolytic mass adjacent to the left frontal sinus wall and the orbital roof | There was no recurrence of the tumor visible on a CT scan 9 months after the operation | An intraosseous cavernous hemangioma characterized by extended, thin-walled vessels and sinuses lined with a single layer of endothelial cells | There was no recurrence of the tumor visible on a CT scan 9 months after the operation |
| Author | Year | Age, y | Sex | Clinical Features | Size | Location | Radiographic Features | Treatments | Pathologic Features | Outcomes |
|--------|------|--------|-----|-------------------|------|----------|----------------------|------------|---------------------|----------|
| Yu     | 2014 | 59     | M   | A slowly enlarging mass in the lateral orbital rim of the left eye for 1 year; a history of craniofacial defect in the left front bone (6 years earlier) | 3 cm × 2.5 cm | Lateral orbital rim of the left eye | CT: a mass with poorly defined margins, which invaded the neighboring tissue and nearly damaged the bone. MRI: hypointense on T1WI and hyperintense on T2WI; significantly inhomogeneous enhancement | Complete resection | Postoperative histopathological examination confirmed that the mass was cavernous hemangioma | The postoperative period was uneventful at follow-up examination after 1 year |
| Murrone | 2014 | 50     | F   | A slow-growing frontal mass, tender to pressure, with spontaneous pain. No history of trauma | – | Left frontal bone | MRI scan showed a hypointense lesion on the T1WI and a hyperintense lesion on the T2WI. CT scan showed an osteolytic lesion with erosion of the skull bone. | A left frontal craniectomy via linear incision was performed with excision of the frontal lesion and a margin of surrounding uninvolved bone and a cranioplasty | A cavernous hemangioma of the diploe with large, thin-walled, dilated capillary spaces lined by flattened endothelial cells without evidence of malignancy | Her postoperative course was uneventful and a CT scan showed complete resection of the mass with a correct cranioplasty |
| Hsiao  | 2015 | 29     | F   | A painful skull defect found incidentally over the right parietal area. The lesion was soft and with mild dimpling. Intermittent pain had started at least 3 weeks before the initial visit | Right parietal (10 mm × 9 mm) and frontal (8 mm × 9 mm) areas of the skull | Right parietal and frontal areas of the skull | CT scan revealed osteolytic lesions with erosion of the skull bone, whereas MRI showed low signals on T1WI, high signals on T2WI, and heterogeneous enhancing effects on gadolinium-enhanced T1WI | A large craniectomy was performed for the evacuation of the 2 osteolytic lesions | The final histological report confirmed the diagnosis of intraosseous cavernous hemangioma | The patient recovered well. She has been followed up for 4 years with no recurrence |
| Kilani | 2015 | 72     | F   | Headache without neurological disturbances | – | Right parietal convexity | A well-defined, extra-axial right parietal convexity space-occupying lesion isointense on T1WI and hyperintense on T2WI. Intense homogeneous post-contrast enhancement | Surgical removal of the lesion via a right parietal approach | Bone trabeculae widely separated by loose connective tissue enclosing multiple thin-walled vascular spaces lined by endothelial cells, suggesting cavernous hemangioma of the skull | — |

CT = computed tomography, MRI = magnetic resonance imaging, PVA = polyvinyl alcohol, T1WI = T1 weighted imaging, T2WI = T2 weighted imaging.
| Lesion              | Clinical Features                                                                 | Radiographic Appearances                                                                                     | Pathologic Features                                                                 | Treatments                                                                 |
|---------------------|-------------------------------------------------------------------------------------|---------------------------------------------------------------------------------------------------------------|---------------------------------------------------------------------------------------|---------------------------------------------------------------------------|
| Osteoma             | Osteomas are slow growing lesions that are normally completely asymptomatic. A few may be associated with Gardner syndrome | CT: small, well-defined round or oval dense and homogeneous lesions. Homogeneous low signal intensity on T1WI; variable appearance on T2WI; not enhance after gadolinium administration | Compact osteomas: mature lamellar bone with no Haversian canals and no fibrous component. Trabecular osteomas: cancellous trabecular bone with marrow surrounded by a cortical bone margin | Not require surgical treatment unless the location or size of the lesion affects the adjacent structures (orbit, sinus, brain) |
| Myeloma             | Bone pain, deterioration of health or abnormalities on blood or urinary test (eg, high erythrocyte sedimentation rate, anemia) | Multiple small, roundish osteolytic lesions that are relatively uniform in size with sharp and non-sclerotic margins. On MRI, the signal intensity of the lesions is nonspecific; a “salt-and-pepper” appearance or diffuse bone marrow replacement may be noted | Myeloma is a malignant disease of the bone marrow characterized by a monoclonal proliferation of plasma cells | Treatment depends on the stage of the disease. The most common treatments are based on chemotherapy or grafting of hematopoietic cells |
| Skull metastases    | Usually secondary to breast, lung, prostate, kidney, and thyroid cancer; generally asymptomatic; may be revealed by a painful swelling | Mostly multiple, well-circumscribed osteolytic lesions, which generally extend into the adjacent soft tissues. Usually homogeneously enhanced on enhanced MRI, but heterogeneous enhancement, peripheral ring enhancement, or lack of enhancement (sclerotic lesions) can be observed | As same as the primary tumor | Surgical treatment may be possible when there is only 1 metastasis, especially if without any neoplastic context. External radiotherapy is another alternative |
| Intracranial meningioma | Predominantly seen in women in the fifth and sixth decades of life and often revealed by painless and expanded swelling | CT: osteosclerotic lesion with destructive irregular and spiculated borders. Low signal intensity on T1WI; variable signal intensity on T2WI; not enhance. Meningeal enhancement is rare and is explained by adjacent dural irritation or invasion, but the center of the tumor growth is outside the dura | Mostly solid tumors with complete capsule and rich vascular supply, usually accompanied with calcification; a few with necrosis, cyst degeneration, hemorrhage | Surgical resection of the lesion is required. The therapeutic decision depends on the possibility of resecting the lesion and on the patient’s health |
| Osseous hemangioma   | Principally located in frontal and parietal bones; discovered incidentally by pain, swelling, facial paralysis, or hearing loss; 3 times more frequent in female during the 40–50th decades | CT: well-circumscribed intradiploic osteolytic lesion; enhanced homogeneously after contrast injection. MRI: isointense on T1WI and hyperintense on T2WI | Mostly are cavernous and contain dilated blood vessels separated by fibrous septa; rarely contain capillaries; often solitary | Mainly is surgery. Embolization may be considered before surgery. Radiotherapy stops tumor growth but cannot reduce its volume |
| Lesion                     | Clinical Features                                                                 | Radiographic Appearances                                      | Pathologic Features                                                                                   | Treatments                                                                                     |
|---------------------------|------------------------------------------------------------------------------------|----------------------------------------------------------------|------------------------------------------------------------------------------------------------------|-----------------------------------------------------------------------------------------------|
| Langerhans histiocytosis  | Clinical features are variable, from asymptomatic lesions to painful swellings    | CT: unequal involvement of the inner and outer tables; appearance of having bevelled edges. The lesion center may contain a sequestrum, representing residual intact bone. MRI: usually strongly enhance after gadolinium administration. | Heterogeneous collections of Langerhans cells with eosinophils, neutrophils, small lymphocytes, and histiocytes. Eosinophilic abscesses may be present demonstrating central necrosis. | Single lesions: conservative treatment (surveillance or systemic corticosteroids). More diffuse or aggressive forms: surgical excision, radiotherapy, and chemotherapy. |
| Epidermoid and dermoid cysts | Painless subcutaneous swelling; discovered mainly during the third and fourth decades; predominantly occur laterally in the parietal or frontal bone | CT: well-demarcated osteolytic lesions with sclerotic borders; may tend to expand into both the inner and outer tables; homogeneously hypodense. MRI: fluidlike signal intensity on T1WI and T2WI and high signal intensity on DWI; usually do not enhance. | Epidermoid cysts, lined with thin squamous epithelium, contain remnants of cholesterol and keratin. | Treatment of these cystic lesions is surgical, usually without recurrence. |
| Aneurysmal bone cyst      | Mainly in children and adolescents; may be secondary to other underlying lesions like fibrous dysplasia, chondroblastoma and osteosarcoma | Sharply defined expanded osteolytic lesion with thin sclerotic borders, although the tables appear disrupted when expansion is significant. | Composed of numerous blood-filled spaces of variable size separated by connective tissue. | The traditional treatment is complete surgical excision. |
| Fibrous dysplasia         | Usually painless; may be revealed by an enlarging mass with symptoms resulting from mass effect; preferentially affects the frontal and temporal bones and may cross the sutures | Mainly affects frontal and temporal bones and may cross sutures; hypointense on T1WI but are sometimes isointense if fibrous tissue is rich. Signal intensity on T2WI is heterogeneous and depends on the fibrous tissue density, intrasoselal cellularity, and hemorrhagic or cystic rearrangements. | Abnormal differentiation and maturation of osteoblasts, with progressive replacement of normal bone by immature woven bone. | Medical treatment is based on bisphosphonates. Surgical decompression is considered in cases of severe mass effect. |
| Paget disease             | Abnormal and excessive remodeling of 1 or more bones in patients typically older than 55 years; particularly affect the frontal and occipital bones; usually asymptomatic and is frequently discovered incidentally | CT: homogeneous enlargement of the skull vault and thickening of the tables and of the trabecular bone. MRI: initially, high signal intensity on T2WI and strong enhancement due to bone resorption; later, fatty bone marrow develops with hypointense thickened trabeculae and surrounded by thick hypointense tables. | Characteristic jigsaw puzzle-like/mosaic pattern. | Treatment relies mainly on bisphosphonates. |
are more inclined to extend externally than intracranially. A variety of clinical manifestations may occur depending on the involved sites. Proptosis and impaired vision may appear if the orbit was invaded. Facial nerve paralysis, twitching of oral commissure, pulsatile tinnitus, and hearing loss may occur if temporal bones are involved. Patients may rarely present with an associated epidural hematoma or subarachnoid hemorrhage. We reviewed some previously reported cases of skull PICH since 2000 (Table 2). The commonest clinical feature is a solid swelling in the skull, painful or painless. Some patients may also present with headache or dizziness.

Radiologic evaluation includes plain skull x-rays, CT scan, and magnetic resonance imaging (MRI). CT is an excellent investigation, as it allows detailed characterization of the cortical and trabecular bone to be made. Although the appearance on CT may vary, an expansive lesion with thin borders and intact internal and external skull plates is the most common finding. MRI signal intensity depends on the amount of venous stasis in the lesion and also on the rate of transformation of red marrow into yellow marrow. Although T1WI may give high- or low-intensity signals, water-sensitive sequences, such as T2WI and FLAIR, commonly give high-intensity signals. The CT or MRI features of the cases previously reported are also shown in Table 2.

The differential diagnosis for intradiploic skull masses includes dermoid tumors, metastatic diseases, meningiomas, sarcomas, Langerhans cell histiocytosis, cosinophilic granulomas, fibrous dysplasia, giant cell tumors, aneurysmal bone cysts, osteomas, Paget diseases, and so on. Because the imaging findings are not specific, preoperative diagnosis is difficult and histopathology is essential. A thorough clinical, radiographic, pathologic, and treatment comparison to other entities in the differential diagnosis is seen in Table 3.

The treatment of choice for skull PICH is total resection with an adequate normal bone margin to reduce the risk of bleeding. The bony defect can be reconstructed by virtue of a variety of methods. Relapse is uncommon if sufficient safety margins are achieved. Other treatment options include curettage which can be followed by recanalization and irradiation, which may reduce the tumor volume and has demonstrated symptomatic improvement, but has the risk of radiation-induced carcinoma. Radiotherapy alone can only can prevent the tumor from growing, but it cannot eradicate the lesions. In keeping with the most widely recommended technique, we opted for a craniectomy with total resection, keeping a 0.5-cm safety margin. We then performed cranioplasty with titanium plate.

**CONCLUSIONS**

PICHs of the skull are rare benign lesions of vascular origin, showing so semblable medical imaging findings to many other bone lesions that it is hard to differentiate them. Thus, there hemorrhagic features within operative fields and histopathologic examinations remain as the "gold standards" for diagnostics. Total resection with enough uninvolved bone margins must be attempted. PICH's relapse is rare when this surgery is successful.

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