Posttraumatic Giant Extradural Intradiploic Epidermoid Cysts of Posterior Cranial Fossa: Case Report and Review of the Literature

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We reported a unique case of posttraumatic giant infratentorial extradural intradiploic epidermoid cyst. A 54-year-old male, with a previous history of an open scalp injury and underlying linear skull fracture in the left occipital region in childhood, presented with a painful subcutaneous swelling, which had developed gradually in the same region and moderate headache, nausea, vomiting and cerebellar ataxia. The duration of symptoms on admission was 3 months. Imaging studies revealed occipital bone destruction and giant extradural intradiploic lesion. The preoperative diagnosis was giant infratentorial extradural intradiploic epidermoid cyst. Surgery achieved total removal of the lesion, which was histologically confirmed and the postoperative course was uneventful. To our knowledge, this is the first case of giant infratentorial extradural intradiploic epidermoid cyst with a traumatic etiology described in the literature.

Key Words: Neoplasm∙ Epidermoid cyst ∙ Cranial fossa ∙ Posterior ∙ Trauma ∙ Etiology

INTRODUCTION

Epidermoid cysts are benign, slow growing lesions, representing about 1% of all intracranial tumors. Intracranial epidermoids are subdivided to more frequent intradural and less common extradural subgroups. Extradural epidermoid cysts are intradiploic in approximately 25% of the cases, and predominantly supratentorial in location. Infratentorial intradiploic epidermoids are not rare whereas the giant variants are extremely rare. We report an unique case of posttraumatic giant infratentorial extradural intradiploic epidermoid cyst. To the best of our knowledge, this is the first case of giant infratentorial extradural intradiploic epidermoid cyst with a traumatic etiology described in the literature. Review of the literature is presented relevant to this unusual case along with their epidemiology, clinic, diagnosis, surgical treatment and etiology.

CASE REPORT

A 54-year-old man experienced bicycle accident with an open scalp injury and underlying linear skull fracture in the left occipital region at 5 years of age. The wound had been treated surgically with lavage and sutures. Subsequently, painless subcutaneous swelling gradually developed in the same region. The patient has been well and without any complaints until 3 months before his admission in our clinic. Local inspection exposed a painful subcutaneous swelling, 7 to 12 cm across, in the occipital region, predominantly on the left side. Physical examination revealed headache, nausea, vomiting and cerebellar ataxia. Computed tomography (CT) exposed a giant infratentorial extradural hypodense lesion with extensive occipital bone destruction and substantial mass effect in the posterior cranial fossa. Magnetic resonance imaging revealed the giant infratentorial extradural hypodense lesion with extensive occipital bone destruction and significant compression of the cerebellum, brain stem, fourth ventricle and the left occipital lobe, without corresponding brain edema. The preoperative diagnosis was giant infratentorial extradural epidermoid cyst.

Under a general anesthesia, mass removal was performed starting with “Hockey-stick” skin incision. The exposed occipital bone was widely destroyed, thinner and partially perforated by the tumor. Magnetic resonance imaging revealed the giant infratentorial extradural epidermoid tumor, inhomogeneously hypointense in T1-weighted images and hyperintense in T2-weighted, with an enhancement rim of the thickened dura mater and significant compression of the cerebellum, brain stem, fourth ventricle and the left occipital lobe, without corresponding brain edema. The preoperative diagnosis was giant infratentorial extradural epidermoid cyst.

The histological
examination confirmed an epidermoid cyst and the postoperative course was uneventful.

DISCUSSION

Epidermoid cysts have expansive type of growth and cranial or spinal localization. Cranial epidermoid cysts are relatively rare (0.3-1.8% of all surgically treated craniocerebral tumors), benign and predominantly-intradural lesions. Extramedullary epidermoid cysts represent about 25% of all cranial epidermoids and engage the scalp or the skull. Extradural epidermoids of the skull are also known as intradiploic. The first primary intradiploic epidermoid cyst was reported by Müller in 1838. Ciappetta et al. cited a total of 223 cases of intradiploic extradural epidermoids, reported in the literature by 1990. Some of these epidermoid cysts may attain giant size before they are diagnosed. The giant intradiploic extradural epidermoid cysts are rather uncommon (about 30 described cases) with a supratentorial predilection for the frontal and parietal bones of the skull.

The first case of infratentorial giant intradiploic extradural epidermoid cyst was reported by Rengachary et al. in 1978. To the best of our knowledge, there have been only 8 cases of such epidermoid in the literature, including the presenting case (Table 1).

The rate of epidermoids growing is slow, linear in contrary to the most of the other tumors with their exponential growth. In correspondence with that the age at the onset of complaints in this group (mean age of 55 years, range 24-74 years) was logically higher compared with that of the non-giant intradiploic epidermoid cysts of the skull (mean age of 32 to 38 years). An utter male sex predilection was outlined in the current series (male: female ratio-8:0) in contrast to the data of other reviews of intradiploic epidermoid cysts of the skull.

The duration of symptoms on admission in the studied group was short and range between 1 and 5 months (mean 3 months), which is most likely explained by the slow expansive growth of the cysts.
by the giant size of the lesions and their significant compressive effect. The presence of painful or painless subcutaneous swelling was not a compulsory, but extremely indicative feature of the diagnosis infratentorial giant intradiploic extradural epidermoid cyst\(^6\). Neurological examination was non-uniformly positive depending on the predominant direction of the tumor growth: intra- or extracranial and the grade of the occipital bone destruction. In both cases with neurologically intact patients, the epidermoid cysts widened the space of the posterior cranial fossa\(^5\).

The imaging diagnosis of the infratentorial giant intradiploic extradural epidermoid cysts does not represent a challenge. X-ray films of the skull and their radiation burden must be avoided, because the lytic occipital bone defect revealed by them is clearly visualized by CT, which in addition demonstrated the giant hypodense lesion and sometimes post-contrast rim enhancement of the thickened dura mater. Regardless that the surgery could be performed on only based on CT scans\(^6\), if available MR imaging should be obligatory in the preoperative investigations, because of its detailed imaging information\(^1\). MR imaging reveal giant extradural lesion inhomogeneously hypointense in T1-weighted and hyperintense in T2-weighted images, with a post-gadolinium dural enhancement over the cerebellum. Magnetic resonance angiography was never performed in the series, but could substitute the conventional angiography with its hazards for the patients, in order to evaluate the grade of compression of the neighbouring venous sinuses.

The differential diagnosis of the infratentorial giant intradiploic extradural epidermoid cysts is quite limited, including dermoid cysts, eosinophilic granulomas, hemangiomas and in some cases-large arachnoid cysts\(^7\) and is easy to be solved.

Despite the huge size of the infratentorial giant intradiploic extradural epidermoid cysts the golden standard of the surgery is the total removal of the tumor with its capsule and preserving the integrity of the dura and its venous sinuses\(^1,2,5,7,8,10\). Neuro-navigation could be useful in the cases with a predominantly intracranial growing and supposed invasion of the dural venous sinuses in order to limit the extent of craniectomy and to preserve the sinuses integrity\(^4\). The total removal of the giant epidermoid cysts leads to permanent cure. Cranioplasty may be needed, when there is a large bony defect. In the series, the etiology of epidermoids was congenital sequestration of ectodermal cells within the cranial bones between the third and fifth embryonic week\(^1,2,5,7,8,10,12\), except in the presented case where the inclusion of ectodermal cells thought to be occurred at the time of trauma.

**CONCLUSION**

We report an unique case of posttraumatic giant infratentori-
| No. | Authors          | Age (yrs) | Sex | Local state                      | Neurological state                  | Duration of symptoms on admission (mths) | Imaging diagnosis                                                                 | Surgery | Supposed aetiology |
|-----|------------------|-----------|-----|----------------------------------|-------------------------------------|-----------------------------------------|------------------------------------------|---------|-------------------|
| 1   | Rengachary et al. | 62        | M   | Negative                         | Headache, nausea, vomiting, intermittent choking spells and nocturnal stridor | Negative                               | Lytic lesion of the occipital bone with expansion and thinning of the inner and outer tables | Total removal | Congenital        |
| 2   | Rubin et al.     | 27        | M   | Negative                         | Headache, occasional diplopia, papilloedema | 2                                 | Isodense, calcification, rim enhancement | Total removal | Congenital        |
| 3   | Guridi et al.    | 47        | M   | Subcutaneous swelling            | Negative                            | 1                                 | Homogeneous radiolucency, lytic occipital bone defect, sharply defined sclerotic borders | Total removal | Congenital        |
| 4   | Jaiswal et al.   | 40        | M   | Subcutaneous swelling            | Cerebellar ataxia                    | Negative                               | Lytic occipital bone defect              | Total removal | Congenital        |
| 5   | Maiuri et al.    | 60        | M   | Subcutaneous bone swelling, local pain | Negative                            | 5                                 | ND                                       | Hyperintense inhomogeneously Enhancing rim | Total removal | Congenital        |
| 6   | Borha et al.     | 73        | M   | Subcutaneous bone swelling, local pain | Headache, cerebellar ataxia, dysmetria | 4                                 | ND                                       | Hyperintense inhomogeneously              | Total removal | Congenital        |
| 7   | Alberione et al. | 74        | M   | Negative                         | Cerebellar ataxia, dysmetria         | Negative                               | Hyperintense calcification, rim enhancement | Total removal | Congenital        |
| 8   | Enchev et al.    | 54        | M   | Subcutaneous swelling, local pain | Headache, nausea, vomiting, cerebellar ataxia | 3                                 | Hyperintense calcification, rim enhancement | Total removal | Trauma            |

CT: computed tomography, NM: not mentioned by the authors, m: male, MRI: magnetic resonance imagining, mths: months, ND: not done, TR: total removal, yrs: years
al extradural intradiploic epidermoid cyst with an established traumatic etiology. The infratentorial giant extradural intradiploic epidermoid cysts are exceptionally rare, extremely slow growing, benign lesions, exclusively in males, with typical X-ray, CT and MRI findings and characteristic dormant clinical course. The non-complicated total removal of these lesions is associated with a good long-term prognosis with permanent cure and lack of recurrence at the follow-up. Caution should be paid in male patients, with an open scalp injury and underlying linear skull fracture in the occipital region and MRI-based follow-up is recommended annually for several years after the trauma, aiming in earlier diagnosis and timely surgical treatment of eventual posttraumatic infratentorial extradural intradiploic epidermoid cysts.

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