A triple Cranial Vault lesion of Hyperostosis Frontalis Interna, Osteoma and Meningioma in a Cadaveric Skull

Tsoucalas G, Siozopoulos A, Vasilopoulos A *, Thomaidis V, Fiska A.

Anatomy Department, Democritus University of Thrace, Alexandroupolis, Greece.

ABSTRACT

Cranial vault lesions are rare and may alter skull figure. Among them, osteomas are the most frequent entities found on burial remains. We present a skull (dry bone) which was unearth in the cemetery of Serres city in Northern Greece and was then donated to the Anatomy Department of the Democritus University of Thrace. The specimen presents a triple lesion and an odd figure, a reason which may explain why it was neglected for a long time period as an unwanted remain of a probably stigmatized person. Radiology (computed tomography) revealed a peculiar combination of a meningioma, an osteoma and a hyperostosis frontalis interna.

KEY WORDS: Cranial Radiology, Meningioma en plaque, Hyperostosis.

INTRODUCTION

Although Skull vault has a limited spectrum of disease that lies between the fields of neurological and musculoskeletal biopathology, it presents a cluster of some rather rare pathological entities, usually with non-specific characteristics. Cranial vault is among human’s most conceivable anatomical regions, formed by the flat bones of the skull. However, unexpectedly it is an area where some benign entities may mimic malignancy, mainly when analyzed by using classical bone-tumor criteria for diagnosis. An incidental finding in radiology, combined with the lack in most cases of clear clinical information, or a non-specific appearance of the lesion, all may further complicate characterization [1-2]. Cranial imaging with radiological examination and in some cases functional imaging techniques, alongside with pathology and clinical image are the methods which testify diagnosis [2-3].

Among the most common intracranial neoplasms in adults we encounter meningiomas. A 4.5% of the cases present a hyperostosis [4], a well-defined relationship firstly described by French physicians Édouard Brissaud (1852-1909) [Figure 1] and Pierre Lareboullet in 1903 [5]. As the bone thickens, tumor infiltration may be found in the medullary spaces. However, only theories exist to explain etiology of hyperostosis in meningiomas [4]. Though meningiomas are histologically classified as benign tumors, treatment and prognosis still remain uncertain and surgical interventions are often complicated due to high variability of presentation and type [6]. Osteomas, firstly described by Venetian physician Giovanni Domenico Santorini (1681-1735) and later on by Italian Anatomist Giovanni Battista Morgagni (1682-1771) [Figure 1] located in skull vault are rare and consist less than 1% of benign bone lesions [7]. Cases presenting osteoma and
meningioma in adults are even rarer [8], while osteomas are speculated to be responsible for local hyperostosis frontalis interna [9] and may trigger the formation and growth of meningiomas [8].

A skull of unknown origin, age and sex with deformities [Figure 2] was unearthed among the unwanted Christian burial remains of the cemetery of Serres in Northern Greece and was then donated to the Laboratory of Anatomy of the Medical School of Democritus University in Thrace. The skull was thoroughly cleansed, studied and underwent through radiology examination, presenting an extremely rare triple vitiation of cranial lesions. We present a cranium with a Hyperostosis Frontalis Interna, a large Osteoma and a Meningioma, all in the cranial vault region.

The bony specimen: Study of the skull specimen, observation of the shape of the supra-orbital ridges, mastoid process, the lower mandible ramus, the cranial sutures and the frontal cranial view (round shape of eyes, v shaped mandible shape from underside), may presume that this is a skull of a female of about 60 years of age.

Radiology, X-ray and computed tomography (CT) scans, revealed a triplet of uncommon malformations, which in combination creates a unique case. The left frontal sinus is occupied by a large osteoma of a mixed type consisting of a Haversian System (osteon) and small islands of trabecular bone [Figure 3], while a Hyperostosis Frontalis Interna (differential diagnosis: Morgagni-Stewart-Morel Syndrome, metastases ?) [Figure 4] is apparent in the frontal bone. Furthermore, a meningioma of the cranial vault causes an extensive hyperostosis mainly in the left parietal area which lies adjacent to the outer table of diploe being normal in thickness (pariosteal type) extended from the coronal to the lambdoid suture [Figure 5].

Table 1: Most common skull vault lesions and imaging characteristics

| N | Disease                        | Characteristics                                                                 | This case |
|---|--------------------------------|---------------------------------------------------------------------------------|----------|
| 1 | Osteoma                        | Progressive enlargement, small, well-defined round or oval lesion that is usually dense and homogenous | ●        |
| 2 | Intraosseous meningioma        | Osteosclerotic lesion, which is frequently associated with destructive irregular and spiculated borders | ●        |
| 3 | Osseous hemangioma             | Well-circumscribed round or oval osteolytic lesion, peripheral sclerosis may be associated |          |
| 4 | Giant cell tumor               | Solid or mixed solid-cystic mass with bone expansion, highly vascularized        |          |
| 5 | Osteosarcoma                   | Osteoblastic, highly destructive, aggressive lesion with periosteal reaction, characteristic "cloud-like" osteoid matrix |          |
| 6 | Fibrous dysplasia              | Focal cystic enlargement of the outer table, characteristic ground-glass matrix appearance, intralesional calcifications |          |
| 7 | Epidermoid and dermoid cysts  | Well-demarcated osteolytic lesions with sclerotic border, homogeneously hypodense, fluidlike signal intensity |          |
| 8 | Paget disease                  | Osteolytic activity producing circumscribed osteoporosis, osteoblastic phase which includes homogeneous enlargement, thickening of the tables and of the trabecular bone, focal diploic dense lesions mimicking enostoses |          |
| 9 | Hyperostosis Frontalis         | Thickening of the frontal bone of the skull                                   | ●        |
| 10| Eosinophilic granuloma         | Well-defined osteolytic lesion, with sclerotic or non-sclerotic margins, and possible periosteal reaction, endosteal scalloping (or erosions) and slight expansion beyond inner and outer tables, central "button sequestrum" |          |
| 11| Myeloma                        | Multiple small, roundish exclusively osteolytic lesions with sharp and nonsclerotic margins |          |
| 12| Lymphoma                       | Small-round cell tumors, can be lytic, sclerotic, or mixed                      |          |
| 13| Renal osteodystrophy           | Diffuse "salt-and-pepper" pattern                                             |          |
| 14| Thalassemia                    | Characteristic "hair-on-end skull"                                            |          |
| 15| Osteopetrosis                  | Diffuse thickening and sclerosis resulting in typical appearance of "bone within a bone" |          |
| 16| Osteitis and osteomyelitis     | Accumulation of pus within the medullary cavity, leading to vascular congestion, formation of reactive bone and granulation tissue around intramedullary pus |          |
| 17| Amyloidosis                    | Tumor-like deposition                                                          |          |
| 18| Traumas                        | Various                                                                        |          |
| 19| Others malignancies            | Tumor-like, ulcer, pigmentation, various                                        |          |
| 20| Metastasis                     | Multiple, well-circumscribed osteolytic lesions, various,                       |          |
Fig. 1: Pioneers on the cranial malformations (from left to right) French physician Édouard Brissaud (1852-1909) Italian anatomist Giovanni Domenico Santorini (1681-1737) and Italian anatomist Giovanni Battista Morgagni (1682-1771).

Fig. 2: The skull of Serres as was unearthed and donated to the Anatomy Department of the Democritus University of Thrace.

Fig. 3: Plain X-ray in frontal and lateral projection (A, B) and CT scan reformations in axial, sagittal and coronal planes (C, D, E) shows a large osteoma (asterisk) of mixed type with Haversian system (arrowhead) and small islands of trabecular bone (arrows) which occupies left frontal sinus.

Fig. 4: CT scan reformations in axial, sagittal and coronal plane (A, B, C) and 3D volume rendering (D). Significant overgrowth of the inner table of the frontal bone (arrows) due to hyperostosis frontalis interna.
DISCUSSION

Meningiomas occur on the sphenoid wing or cranial convexity, observed as tumors usually near the sutures [4,10]. They present a male to female ratio 1:1.1 with an age range from 7 months up to 82 years, recording an incidence between 10% and 20% in the general population. Although most are histologically benign presenting a slow-growing evolution, malignant cases do occur [11]. Hyperostosis is a common imaging characteristic of most meningiomas, a feature which predominates in meningiomas en plaque (MEP). Computed tomography (CT) usually reveals erosions, sclerosis and the hyperostosis itself, while a subdural plaque of ossification may be seen separated from the sclerotic and/or hyperostotic bone by a linear translucency corresponding to dura mater [12]. Meningiomas on radiology may be hyperostotic, osteolytic or mixed, while areas of hypostosis have been noted too [13]. Most likely hyperostosis and various bony changes in meningiomas represent nothing more than a local reaction (preceding trauma, irritation and/or invention by the tumor, stimulation of osteoblasts, vascular disturbances?) [14]. Wide surgical excision seems to be the choice of treatment. However, 22% of the cases experience recurrence and 26% present atypical malignant changes and metastases mainly towards the whole skull and the spine [15].

Osteomas are benign mesenchymal osteoblastic tumors which present progressive enlargement. Cranial vault is the main area of manifestation, with the temporal bone to be rarely affected and the frontoethmoidal region to be the most common [8]. The most frequent site is the frontal sinus, occurring in 71.8% of the cases [16]. Injuries, inflammation, genetic defects and developmental disorders are the factors of an uncertain aetiology, factors which may trigger osteomas. Osteomas have a slower rate of progression in comparison with meningiomas and in some case recurrence of the tumor may occur [8]. Histologically they may be divided into ivory, mature or mixed. Ivory type osteomas are composed if dense bone without Harvesian systems, while mature ones of cancellous bony material with Harvesian systems. There is a male predilection, with an occurrence in individuals of 30 up to 60 years of age. In burial and archaeological remains, osteomas, are the most commonly found tumors, claimed as easy to be identified [16]. Osteomas may co-exist or trigger, or even mimic meningiomas [7-8] and may be the reason for hyperostosis frontalis interna to occur [9].

Hyperostosis frontalis interna is a pathological entity with an unknown aetiology and in
most cases an incidental finding on skull radiology. It is a condition characterized by excess bone growth and manifests on the inner table of the frontal bone, occasionally extending onto the temporal and parietal, as well as to the occipital bones. It occurs in 5% to 12% of the general population, primarily in females of the age group 40 to 60 after significant hormonal imbalance, such as menopause. The male to female ratio is 1:9 [17-19]. Although some believe that there is a connection with osteomas [9], others mention no relationship [19].

Hyperostosis frontalis interna and meningiomas demonstrate a female predisposition, in accordance to the specimen presented [11, 17-19]. Various studies have recorded combinations of two cranial lesions [4, 7-9], while the majority describe one and/or metastasis [1, 11-12]. Nonetheless, to our knowledge, there are no triple lesion cases reported, a fact which at least means rarity of the case. Meningioma presents an extended hyperostosis of the cranial convexity, diffused in a area between and in proximity with the coronal and lambdoid sutures, as expected [4]. Hormonal imbalance in many cases of the female gender, as the simultaneous presence of other benign masses seams to justify hyperostosis frontalis interna [9, 17-19]. Age range for the appearance of all three lesions is in accordance to the bony specimen in study [11, 16]. There has been reports that patients with hyperostosis frontalis interna exhibit neuropsychiatric symptomatology [20]. The same apply to the meningiomas too, while similar symptoms like vertigo and nystagmus may be produced by osteomas [11,19].

This fact alone, combined with the skull deformities of the presented case, could create a figure and personality not easily tolerated among the rural population of the late 19th century in Northern Greece. Thus, we may speculate the reason why the skull had been neglected among the unclaimed burial remains.

CONCLUSION
Cranial vault bony lesions are entities which may co-exist, create a diagnosis confusion and trigger one the other. Skeleton study and radiology may produce a possible diagnosis. Rare cases may enrich our knowledge and in turn introduce further debate.

Conflicts of Interests: None

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