A Case Report of Osteochondroma of the Frontotemporosphenoidal Suture

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Osteochondroma, also known as osteocartilaginous exostosis, is the most frequent benign bone tumour of the skeletal system.
Despite its preference for long bones (tibia and femur), osteochondroma may occur in some short bones developing endochondral ossification.
Seventy-five percent of the patients present only with a single lesion, whereas 25% have multiple lesions; this last clinical condition, defined as osteochondromatosis (disorder of autosomal dominant inheritance) shows a high risk of malignant transformation (about 11%).
In the craniofacial area this tumour is very rare. The sites of predilection are the coronoid process and mandibular condyle, even though osteochondromas arising from the base of the skull, maxillary sinus and zygomatic arch have been previously described. However, an osteochondroma originating from the frontotemporosphenoidal suture has not been reported before in the literature. We present a unique case of osteochondroma of the frontotemporosphenoidal suture.
Moreover, the relevant international literature has been reviewed and all diagnostic and surgical matters have been discussed.

Key Words: Osteochondroma, Osteocartilaginous exostosis, Benign bone tumour

Osteochondroma, also described as osteocartilaginous exostosis, is the most common benign tumour of the axial skeleton and represents between 35% and 50% of all the benign bone neoplasms (1-3).

This neoplasm is more frequent in males rather than in females (Ratio M/F 1.6:1), and it mainly affects people between the age of 20-30 years (4-6).

Such a neoformation appears clinically as a growing osteocartilaginous protuberance originating from the external cortical surface of the bony segment affected.

Osteochondroma is considered as a cartilaginous tumour, because its bony mass is produced by progressive endochondral ossification of its growing cartilaginous cap (7).

Seventy-five percent of the patients presented a single lesion, whereas 25% had multiple lesions (4); this last clinical condition, defined as osteochondromatosis is recognized as disorder of autosomal dominant inheritance (8).

The difference between single localization and osteochondromatosis is really important in order to distinguish the different risk of sarcomatous transformation: about 1% for a single osteochondroma versus 11% for ostechondromatosis (9).

The most common sites of occurrence are long bones of the lower extremity, such as distal methaphysis of the femur and the one proximal of the tibia (8); nevertheless, other anatomic localizations of osteochondroma include short bones with enchondral ossification such as scapula, pelvis, and spine, synovial joints, and more rarely soft tissue (3, 10-12).

On the craniofacial district the incidence of osteochondroma is extremely low. Osteochondromas of the base of skull, maxillary sinus, zygomatic arch have been reported in literature even though such lesions arise with a higher rate either from the coronoid process or from the mandibular condyle (13-17).

In 1985, Barnes (18), by reviewing international literature, observed the anatomic distribution of 63 soli-

Table I - Anatomic Distribution of 63 osteochondromas arising from head and neck (Barnes, 1985)(18)

Location	No. of Cases
Mandibole	39 (62%)
n Coronoid Process	20
n Condyle	15
n Not specified	2
n Canine Area	1
n Symphysis	1
Cervical Vertebraes	14 (22%)
Base of Skull	6 (10%)
Zygomatic Arch	2 (3%)
Maxillary Sinus	1 (1.6%)
Larynx	1 (1.6%)

tary osseous, osteochondromas of the head and neck; in 62% of cases the mandible was the site most involved (Table I).

In 54% of the cases the local sites were in coronoid process, 41% in mandibular condyle, and 3% in mandibular symphysis (19).

This particular distribution is due to embryology, considering the enchondral ossification growing of the mandibular coronoid process, condyle and symphysis (7).

In a recent study, Shankly et al. (20) have reviewed the British literature by presenting 96 cases of splanchnocranium osteochondromas (Table II). After this report, only 8 other cases have been reported (17, 21-24).

In this article we describe a case of osteochondroma of the frontotemporosphenoidal suture, never reported before in literature.

Case Report

A 34-year-old female was referred to us with a bony protuberance on the right frontal area. The patient

Table II - Osteochondromas of the maxillofacial district (Shankly et al.) (20)

Location	No. of Cases	Gender	Age
Mandibole		Male-Female	
n Condyle	33	15 - 18	19 - 69
n Coronoid Process	30	21 - 9	10 - 73
n Angle	1	0 - 1	34
n Symphysis	1	0 - 1	28
n Alveolare Crest	18	4 - 14	44 - 83
Tongue	8	4 - 4	9 - 73
Zygomatic Arch	2	0 - 2	11 - 17
Maxillary Tuberosity	1	0 - 1	50
Nasal Septum	1	1 - 0	32
Maxilla	1	0 - 1	35
Total	**96**	**45 - 51**	**9 - 83**

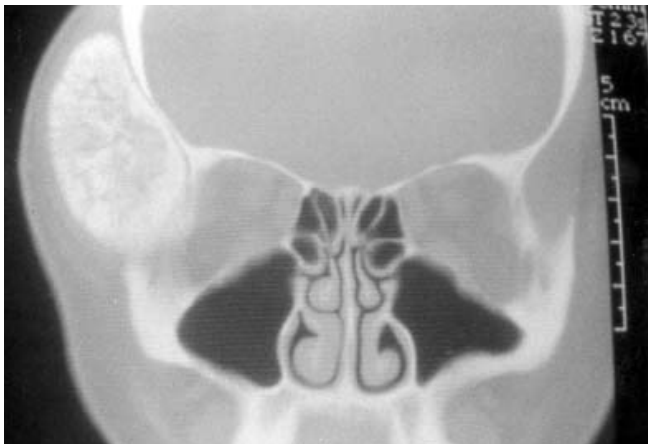

Fig. 1 - Preoperative CT in coronal section. Presence of the tumour at the pterion

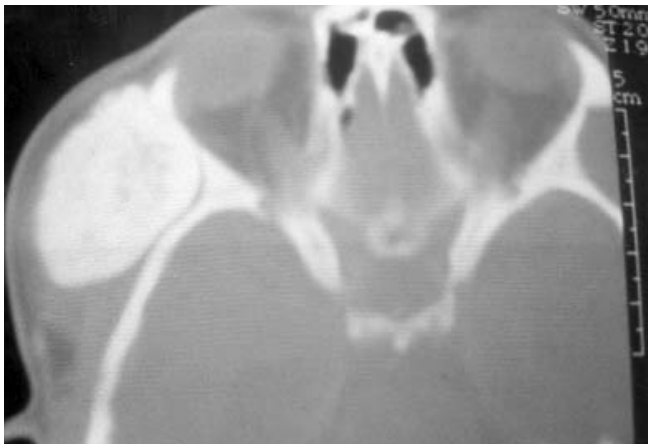

Fig. 2 - Preoperative CT in axial section showed enhancing tumour.

explained to us that, 15 years earlier, she had noted the presence of a limited protuberance in the same area. And, six years later, because of the gradually growing bony mass, she underwent elsewhere the first operation for tumour removal under general anaesthesia.

The histopathological diagnosis was osteochondroma. Only one year following this procedure, the patient noted a recurrence in the area surgically treated but, she did not undergo further investigations because the mass was asymptomatic despite the progressive volume augmentation.

Seven years after the first operation, the patient, however, decided to remove the lesion because she was concerned about the cosmetic appearance. We performed some pre-operative investigations such as total body scintigraphy and Skull-Face CT scan which were able to identify and delineate the osteochondroma and show its localization on the right frontal orbital area. (Fig. 1, 2)

On clinical examination an asymmetry of the third superior of the face was evident due to the presence of a large bony mass on the right frontoparietal district. The mass was palpated as a bony, hard lesion and measured 5 cm in length and 6 cm in diameter.

CT scan revealed a pedunculated bony mass on the right frontosupraorbital area; such lesion appeared as part of the frontal bone, protruding posteriorly as an exophytic component ending at the lateral border of the frontal bone and the lateral orbital wall, resulting much thinner.

According to these findings the diagnosis was recurrent osteochondroma of the frontotemporosphenoidal suture. For this reason the patient was operated under general anaesthesia. A hemicoronal incision was made to allow the exposure of temporalis fossa and temporalis muscle; therefore, through a subfascial approach the right zygomatic arch was identified. The temporalis muscle was dissected and inferiorly rotated, thus exposing a large mass (5 cm in diameter) which arose from the pterion. The whole tumour was removed preserving dura mater which was intact. In this case, the resulting defect was reconstructed by enhancing the temporalis muscle flap (Fig. 3).

Histology report confirmed the diagnosis of Osteochondroma.

The tumour removal was complete as documented with CT scan performed postoperatively (Fig. 4).

Discussion

The World Health Organization recognizes osteochondroma as osteocartilaginous lesion protruding from the external cortex of the bone affected (25).

Typical histology features are represented from chondrocytes, presenting normal characters, placed in parallel columns with the presence of lacunar areas similar to those localized on the cartilaginous epiphysis; in the central zone, there are regular bony trabeculas developing through endochondral ossification (26).

The exostosis is covered by periosteum, which is connected with the one of the contiguous bone (8).

Osteochondroma pathogenesis is a well debated topic and thus several theories have been advocated:

1) such pathology originates from those bones which present with endochondral ossification (2);

2) herniation of the cartilage (27);

3) multiple mechanical traumas at tendon insertion level might cause a proliferation of pluripotent cells

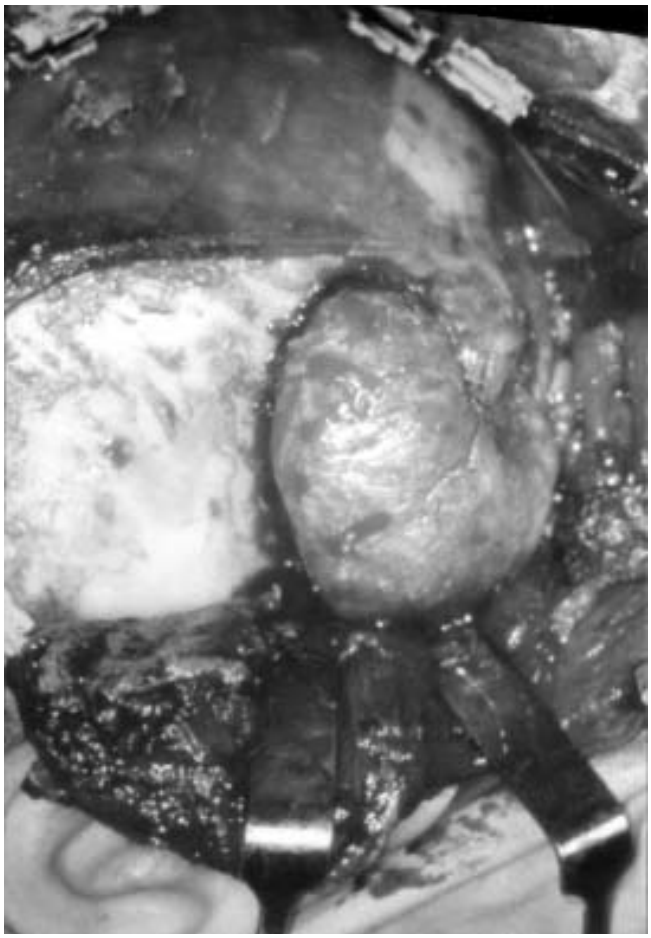

Fig. 3 - Intraoperative details.

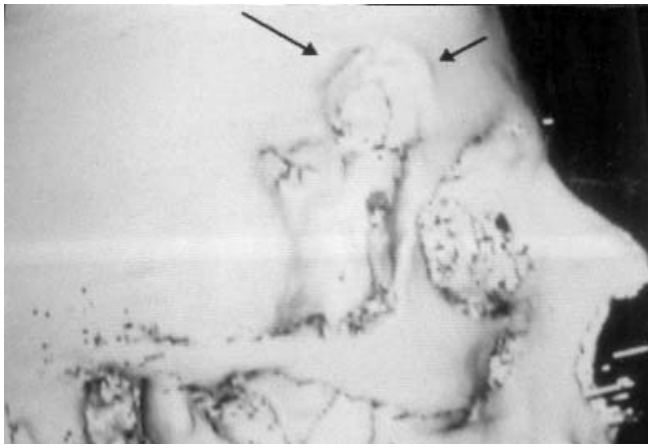

Fig. 4 - 3D CT after tumour removal. The mass was removed at the zone of bony depression *(arrows)*.

resulting in a chondroblasts transformation (28);

4) pluripotent cells metaplasia of the periosteum leading to endochondral ossification (29);

5) activation of cartilaginous cells arising from skull synchondrosis (14-30);

6) traumatisms (31).

According to the second theory, Hwang and Peck demonstred that by inducing a lack of periosteum on the proximal methaphysis in immature rats, a higher incidence of osteochondromas occurred during their growth (32).

Any minimal and frequent injury of the coronoid process and of the condyle, however, seems to validate the third theory.

Whereas the fourth, fifth and sixth theories provide a series of explanations and causes regarding our case of osteochondroma described in this report.

According to our opinion, considering all the different anatomic sites, at least 2 or more of the mentioned theories might be involved in the genesis and development of the neoplasia.

On the basis of anatomic site, there is a wide range of symptoms such as a swelling and a protuberance with a progressive facial asymmetry, articular pain and dysfunctions, malocclusion (17).

Traditional radiographic investigations often provide a nearly satisfactory support in the diagnosis, but modern diagnostic procedures such as CT scan and MRI permit a more detailed and meticulous visualization of the lesion and also the possibility to preoperatively plan the surgical procedure.

Moreover, in this case report, CT scan allowed us to recognize the neoplasm as recurrence and not as malignant transformation; this finding was confirmed from the histology which can be considered as unique reliable procedure to guarantee the definitive diagnosis and to exclude a possible malignant conversion (30).

Osteochondroma can be considered a benign tumour which does not recur if completely excised (23). In literature only one case of recurrence in the craniofacial district has been described (1). Therefore, it is possible to perform in one stage either the tumour removal and the surgical reconstruction if required.

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