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Case Report

Ethmoid-Nasal Phosphaturic Mesenchymal Tumour

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Abstract

Phosphaturic Mesenchymal Tumour (PMT) is a very rare benign neoplasm. Weinder and Santa Cruz first used the acronym PMT in 1987, due to the histological polymorphism shown by the neoplasm. About 95% of PMT involves bones and only 5% involves ENT districts. The clinical picture of PMT is characterized by TIO or tumour-induced osteomalacia, which causes bone pain, muscular weakness and pathological fractures and causes symptoms that can vary, depending on the location. The knowledge of the biological process of this rare neoplasm is therefore crucial to treat it. Due to its local aggressiveness, there is indication for a surgical resection with clean surgical margins. The surgical treatment firstly aims at reducing local aggressiveness, limiting the invasion to the adjacent structures, such as the base of the skull, orbit or palate and, as a consequence, the paraneoplastic syndrome induced by the tumour with its sequences. Most described PMT cases are benign, even though some episodes of malignant transformation with distance metastasis were registered.

Keywords: Phosphaturic mesenchymal tumour; Phosphaturia; Head and neck neoplasm; Sinonasal; TIO; paraneoplastic syndrome. The cases linked to head and neck district were stratified per location and the cases that involved other sections were not considered.

Clinical Case

Male patient, 53 years old, had been showing nasal obstruction for six months, otitis treated with inhaled antibiotics and crenotherapy with no results. He also reported paresthesia on left hard palate [1]. He underwent rhinoscopy with optical fibres showing evidence of a neoformation of the right nasal concha, basis right of the nasal cavity with subtotal occlusion of the right posterior nasal aperture which impeded to see the Eustachian tube orifice. A CT with contrast medium confirmed the presence of a neoformation with partial involvement of the right pterygopalatine fossa, delimited by a confining bone. As the neoformation was located, the patient underwent an endoscopic transnasal surgical removal of the neoformation. The histological result initially showed the neoformation as a pleomorphic adenoma of the right nasal cavity. After a revision of the slides in another centre a phosphaturic mesenchymal tumour was diagnosed, due to the histological polymorphism characterized by hyalinization with microcystic spaces and a prevalence of chondroid and myxochondroid pattern. Later, the patient underwent a revision of the surgery with enlargement of the surgical margins and with a completely negative histological result for the disease. Moreover the concentration of calcium, phosphatemia and phosphaturia excluded Tumour-Induced Osteomalacia (TIO). Today, after 8 months since the second surgery, the patient appears to be free from the disease.

Discussion

ETM PMTs are extremely rare. The diagnose is usually late due to the presence of osteomalacia or to symptoms linked to a local invasion [3]. Patients affected by paraneoplastic syndrome can show non-specific bone pain, muscular weakness and pathological fractures [2-5]. PMT doesn't show to have any link with sex and emerges to affect a very ample range of patients, from the age of 3 to 73 [2-5]. 95% of PMTs were observed in bones and only 5% in craniofacial districts [2-7]. Among these about 50% of tumours were found in the sinonasal tract [2-8]. In our revision it was observed that 20 cases of PMT reported in literature involve the sinonasal tract, while the rest of them involve other ETM tracts, as mandible, mouth, pharynx, larynx, thyroid and temporal bone [2-8]. From a histological point of view PMT is characterized by spindle-shaped or stellar cells in a myxo-chondroid or myxoid matrix with calcification [2-5]. Osteocytes in PMT are responsible for osteomalacia through the production of Fibroblast Grow Factor 23 (FGF23), which inhibits the transport of the sodium phosphate renal tubules leading to a phosphaturia and a consequent bone demineralization [9,10]. PMT is mostly suspected when a not familial hypophosphatemia is present. Diagnostic work-up must take into consideration the patient's history, an objective systemic exam and a search for localizations in arms and legs and ETM [10]. An otorhinolaryngologist who finds himself in front of a histological PMT must investigate a possible osteomalacia, as most cases see the presence of a paraneoplastic syndrome, and exclude the involvement of the bones [11]. In the series displayed in chart 1 it is possible to observe that only 6 patients didn't present TIO. In an extensive revision carried out by Folpe et al. on 109 mesenchymal tumours on the extremities, only 3 cases didn't present TIO [2]. The first line treatment is surgical resection with ample margins, which leads to a normalization of phosphatemia and phosphaturia with an improvement of the mineralization of bones [2]. The persistence of metabolic alterations after surgical resection is predictive of an incomplete surgical resection or a relapse [1]. Surgery appears to be the best choice also for the rare malignant manifestations of PMT, while adjuvant chemotherapeutic treatments haven't been established, yet, due to the small amount of cases [12].

Results

From the revision of literature 405 articles containing PMT cases

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Table 1: Clinical pictures present in literature.

AGE AUTHORS'	LOCATION	S.P.*
Sinonasal PMTs		
Linsey et al. [14] 54F	RHINOPHARYNX	YES
Weidner and Santa Cruz [1] 35F	MAXILLARY SINUS	YES
Papotti et al. [13] 38F	NASAL CAVITIES, MAXILLARY SINUS, ETHMOID	YES
Gonzalez-Compta et al. [11] 69F	ETHMOID, FRONTAL AND MAXILLARY SINUS	YES
Kawai et al. [15] 53F	RIGHT NASAL CAVITY, ETHMOID	YES
Ungari et al. [6] 24M	ETHMOID	YES
Inokuchi et al. [16] 24F	NASAL CAVITIES, MAXILLARY SINUS, ETHMOID, FRONTAL SINUS	YES
Koriyama et al. [17] 41F	MAXILLARITY SINUS	YES
Winters et al. [18] 55F	PREAURICOLAR REGION	NO
Pedrazzoli et al. [19] 37F	RIGHT MAXILLARY SINUS	YES
Shelekhova et al. [20] 70F	NASAL CAVITY FRONTAL SINUS	YES
53F	FRONTAL SINUS	YES
Peterson et al. [21] 33F	MAXILLARY SINUS	YES
Parshwanath et al. [22] 42F		YES
Komínek et al. [23] 53M	FRONTAL SINUS, ETHMOID, NASAL CAVITY	NO
Guglielmi et al. [24] 22M	RHINOPHARYNX, ETHMOID, SPHENOID SINUS	YES
Battoo et al. [25] 34F	NASAL CAVITY, ETHMOID, MAXILLARY SINUS	YES
Deep 4 41M	RHINOPHARYNX	NO
Demetri A. et al. [5] 50F	NASAL CAVITY	YES
**Spinato R, Politi D. et al. 2015 53M	NASAL CAVITY, PTERYGOPALATINE FOSSA	NO
Nonsinonasal head and neck PMTs		
Olefsky et al. [26] 40M	PHARYNX	YES
Shenker and Grekin [27] 55M		YES
Weidner and Santa Cruz [1] 27M		YES
Harvey et al. [28] 32F	THYROID	YES
Yang et al. [29] 31F	PERIMANDIBULAR TISSUES	YES
Reyes-Múgica et al. [30] 9F		YES
Dupond et al. [31] 71M	MANDIBULAR ALVEOLAR PROCESS	YES
Woo et al. [8] 42F	MANDIBLE	YES
Kaylie et al. [32] 46F	TEMPORAL BONE	YES
Uramoto et al. [33] 48M	TONGUE	YES
Yun et al. [34] 71F	BASE OF THE ORAL CAVITY	YES
Savage and Zimmer [35] 73F		YES
Mori et al. [36] 42M	MANDIBULAR	YES
Sidell et al. [12] 24F	LARINGIS	NO
Syed et al. [37] UNKNOWN AGE	TEMPORAL BONE	NO
Luo et al. [38] UNKNOWN AGE	MANDIBLE	YES

*PS: Paraneoplastic Syndrome
**Clinical Study of the Present Article

from 1970 to 2015 were selected.

36 ETM PMT cases were found, among those 20 localized on a nasal-sinusal level and 16 in other locations, such as mandible, base of the mouth, pharynx, larynx, thyroid and temporal bone (Table 1).

Conclusion

PMT in head and neck districts is usually benign and associated to paraneoplastic syndrome characterized by tumour-induced osteomalacia. About half of PMTs are localized in the sinonasal

district. An informed otorhinolaryngologist of this rare case emerges to be a very important factor for the right treatment of these tumours.

Surgery seems to be the best choice being able - with ample margins - to control local aggressiveness and osteomalacia induced by the majority of these tumours.

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