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Case Report

Psammomatous ossifying fibroma of frontal sinus: a case report with review of literature

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ABSTRACT

Psammomatoid Ossifying Fibroma (POF) is a rare, slowly progressive benign tumor of the extragnathic craniofacial bones representing a subgroup of related fibro-osseous lesions with a tendency towards locally aggressive behaviour unique to the nasal cavity, paranasal sinuses, orbit and is often misdiagnosed. Histologically they are benign, but clinically they are locally aggressive. We report a case of a 17 year-old boy who presented with psammamatous ossifying fibroma of the frontal sinus. The location of this tumor made this case unusual.

Keywords: Frontal sinus, Psammomatoid ossifying fibroma, Young adult

INTRODUCTION

Ossifying fibroma is a rare benign tumor of the nasal cavity and paranasal sinus. Psammomatoid Ossifying Fibroma (POF), also known as juvenile ossifying fibroma, is a rare tumor of the extragnathic craniofacial bones, particularly the periorbital, frontal, and ethmoid bones. It is a slowly progressive lesion with a tendency toward locally aggressive behavior, including invasion and destruction of surrounding tissue, bony erosion, and recurrence after surgical excision and is histologically characterized by the presence of spherical ossicles, which are similar to psammoma bodies.[1] In our case the lesion was seen focally infiltrating bone. However, there was no significant nuclear pleomorphism, and mitoses could not be demonstrated. We are presenting a case of psammomatoid ossifying fibroma, due to its unusual location.

CASE REPORT

A 17 year-old male patient presented with a mild forehead swelling of 3 weeks duration accompanied by dizziness. Radiographic examination revealed an expansile well-circumscribed radiolucent lesion surrounded by a thick bony wall. CT examination revealed dilation of the right frontal sinus with ground glass appearance with intact cortical outline. The frontal sinus was communicating with the nasal cavity and a diagnosis of fibrous dysplasia was offered. After hospitalization, a right frontal sinus fenestration and tumor resection plus nasofrontal duct reconstruction was conducted under general anaesthesia.

Gross: Received multiple grey brown to dark brown bits mixed with few bony bits all together measuring 2 cm x 2 cm.

Microscopy revealed proliferative fibrous tissue composed of proliferated oval to spindly cells that often formed small whorls with interspersed multiple foci of concentric calcified spherules reminiscent of psammoma bodies with few intervening bony spicules showing the presence of osteoblastic rimming. Some bony spicules lacked the osteoblastic rimming. The lesion was seen focally infiltrating bone. However, there was no

significant nuclear pleomorphism, and mitoses could not be demonstrated.

Diagnosis of psammomatous ossifying fibroma lesion was given. The postoperative period was uneventful and patient was lost to follow up.

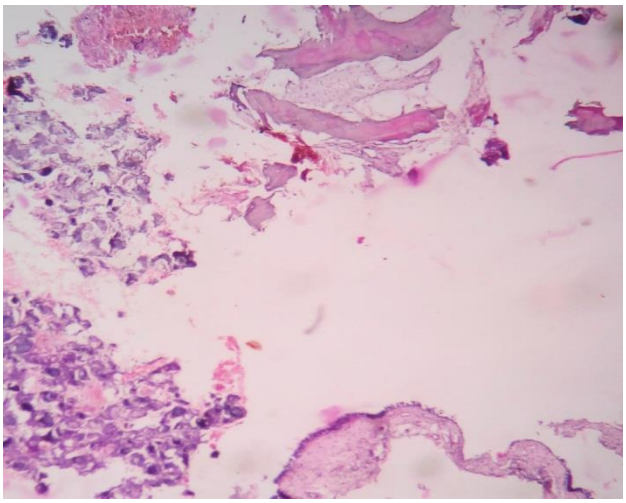

Figure 1: Tumour with surrounding bone spicules showing osteoblastic rimming at places (H&E 40x).

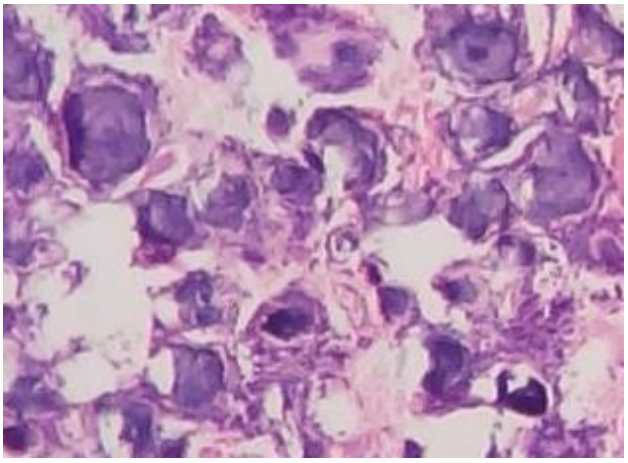

Figure 2: Psammamatous calcification within the tumour (H&E 100x).

DISCUSSION

Gogl initially described POF[2] as "psammomatoid fibroma of the nose and paranasal sinuses in 1949". Margo et al. in 1985,[3] described POF as a distinctive solitary fibro-osseous lesion of young person's that affects the orbit and shows characteristic histologic features. POF has also been reported under the designations of "juvenile ossifying fibroma" with psammoma-like ossicles by Slootweg et al.[4] Within the spectrum of benign fibro-osseous lesions of the jaws, ossifying fibromas represent the neoplastic entities. The precise nature and classification of the juvenile ossifying fibromas has been the source of considerable debate among pathologists, resulting in a confusing proliferation of competing nomenclatures.[5] This controversy is further exacerbated by the fact that this group of tumors are not restricted to paediatric patients with morphologically identical lesions identified in adults. A degree of consensus has been established and most sources accept the existence of juvenile ossifying fibromas which are further subdivided into juvenile Psammomatoid Ossifying Fibromas (JPOF) and Juvenile Trabecular Ossifying Fibromas (JTOF) subtypes. The Ossifying Fibromas (OF) are subdivided into juvenile clinicopathologic and conventional subtypes. On the basis of morphologic features, juvenile ossifying fibromas are further separated into trabecular (JTOF) and psammomatoid (JPOF) variants.[6] The juvenile variants are characterized by distinctive trabecular or psammomatoid matrix production, occurrence in younger patients relative to conventional OF and a predilection for the bones of the paranasal sinuses, the periorbital region and the maxilla.[4,7,8] In contrast, conventional OFs tend to be more common in the mandible. It probably arises from overproduction of the myxofibrous cellular stroma normally involved in the development of the septa in the paranasal sinuses as they enlarge and pneumatize. Hyaline material secreted by these stromal cells ossifies and connective tissue mucin initiates the cystic areas. The myxoid tissue, cystic degeneration, and aneurysmal-bone-cyst-like areas characteristic of some juvenile POF considered as reactive changes were not seen in this lesion.[9] The majority of cases have been reported in children and young adults between 5 and 25 years of age with slight male predominance (1.2:1). The lesion is particularly common in the periorbital, frontal, and ethmoid bones. When seen in children they are a more aggressive variant and are known as juvenile aggressive cemento-ossifying fibromas. Clinical presentation is with a solid mass, arising usually from the mandible (62-89%) or maxilla. Most frequently they arise from the premolar region of the mandible (77%).[10] Our patient presented only with a frontal sinus swelling with mild dizziness though other clinical manifestations of the POF include progressive craniofacial deformities proptosis, visual disturbances, progressive blindness, airway obstruction and headache. Local expansion of the tumor may extend into adjacent structures, such as the paranasal sinuses, nasal cavity, nasopharynx, palate, and cranial cavity. In this case the tumour was small and was restricted to the frontal sinus. Entities considered in the radiographic differential diagnosis of this lesion include fibrous dysplasia and mixed odontogenic tumors. The mixed odontogenic tumors, odontoma and ameloblastic fibro-odontoma also occur in younger patients and present as mixed density mass lesions in the tooth bearing portions of the jaws was not considered in the diagnosis. Fibrous dysplasia also presents as a ground glass radiopacity and was considered in diagnosis .Histologic feature of POF is the presence of numerous small, round ossicles or "psammomatoid" bodies that are embedded in a cellular fibrous stroma. The ossicles are mineralized collagenous foci that vary from small, smoothly contoured round-to-oval patterns to

larger, irregularly shaped patterns, with concentric layering similar to that of psammoma bodies. A prominent marginal osteoid rim surrounds the ossicles. The main histologic differential diagnosis is a primary sinonasal meningioma. The completely extradural location and the partly cystic nature of the present lesion, the young age of the patient, and the EMA negativity of the tumor cells favor the diagnosis of POF over meningioma.

CONCLUSION

Cemento-ossifying fibromas commonly occurring in children are usually well circumscribed masses which expand the underlying bone. They are usually small, but can become large when they arise from the maxilla or paranasal sinuses because there is more room to expand. They usually expand the bone without cortical breach. Surgical excision is the treatment of choice and it usually requires bone grafting or reconstructive surgery.[10,11] Recurrence following complete excision is generally considered to be uncommon.[11,12]

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