Kulak Burun Bogaz Ihtis Derg 2008;18(3):185-187

CASE REPORT

A rare cause of exophthalmos: cemento-ossifying fibroma

Ekzoftalmusun nadir bir nedeni: Semento-ossifiye fibroma

Melda APAYDIN, M.D., Çağlar ÇALLI, M.D.,[1] Bengü GÜNAY YARDIM, M.D.,[2]
Ayşegül SARSILMAZ, M.D., Makbule VARER, M.D., Engin ULUÇ, M.D.

Cemento-ossifying fibroma is a benign fibroosseous lesion that contains fibrous tissue and calcified tissue resembling bone, cementum or both. It is frequently seen in the mandibula and maxilla, but it may rarely affect the ethmoid sinus. In this report, we presented computed tomography findings of an ossifying fibroma of the ethmoid sinus associated with exophthalmos. A 25-year-old woman presented with complaints of exophthalmos, headache, and nasal congestion of six-month history. Physical examination showed a firm mass on the right side of the nasal septum and right-sided exophthalmos. Eye movements, vision, and the fundus were normal. Axial and coronal computed tomography scans showed a well-delineated, round mass, 4x4.5x3 cm in size, in the right ethmoid sinus, extending from the right orbital rim to the right nasal cavity. Near-total excision of the mass was performed by a lateral rhinotomy and medial maxillotomy approach. Based on histologic and radiological findings, the diagnosis was made as ossifying fibroma.

Key Words: Bone neoplasms; ethmoid sinus; exophthalmos/etiology; fibroma, ossifying/diagnosis/pathology; paranasal sinus neoplasms.

Semento-ossifiye fibroma içinde fibröz doku ve kemik, sementum ya da her ikisini andıran kalsifiye doku içeren benign fibro-osseöz bir lezyondur. Sıklıkla mandibula ve maksillayı, nadiren de etmoid sinüsü etkiler. Bu yazıda, etmoid sinüste gelişen ve ekzoftalmus oluşturan ossifiye fibromaya ait bilgisayarlı tomografi bulguları sunuldu. Yirmi beş yaşındaki bir kadın hasta, altı aydır var olan ekzoftalmus, baş ağrısı ve burunda konjesyon yakınmalarıyla başvurdu. Fizik muayenede, nazal septumun sağ tarafında sert bir kitle ve aynı tarafta ekzoftalmus saptandı. Göz hareketleri, görme ve fundus normal bulundu. Aksiyel ve koronal bilgisayarlı tomografi görüntülerinde, sağ etmoid sinüste, sınırları belirgin, yuvarlak ve sağ göz çukuru kenarından sağ nazal kaviteye kadar uzanan, 4x4.5x3 cm büyüklüğünde bir kitle görüldü. Kitle lateral rinotomi ve medial maksillotomi yaklaşımıyla tama yakın çıkarıldı. Histolojik ve radyolojik bulgular ışığında tanı ossifiye fibroma olarak kondu.

Anahtar Sözcükler: Kemik neoplazileri; etmoid sinüs; ekzoftalmus/etyoloji; fibroma, ossifiye/tanı/patoloji; paranazal sinüs neoplazileri.

Cemento-ossifying fibroma (COF) is a benign maxillofacial fibroosseous lesion (FOL) containing fibrous tissue and varying amounts of calcified tissue resembling bone, cementum or both. Fibroosseous lesions of the face and jaw are cemento-ossifying dysplasia, fibrous dysplasia, and cemento-ossifying fibroma.[1,2] Cemento-ossifying fibroma is frequently seen in the mandibula and maxilla, but it may rarely involve the ethmoidal region or orbits. It is also known as ossifying fibroma, juvenile ossifying fibroma, cementifying fibroma, and juvenile active ossifying fibroma.[1-4]

◆ *Departments of Radiology, [1]Otolaryngology, and [2]Pathology, İzmir Atatürk Training and Research Hospital (İzmir Atatürk Eğitim ve Araştırma Hastanesi, Radyoloji Kliniği, [1]Kulak Burun Boğaz Hastalıkları Kliniği, [2]Patoloji Kliniği), İzmir, Turkey.*

◆ *Received - June 5, 2006 (Dergiye geliş tarihi - 5 Hazian 2006). Request for revision - May 1, 2007 (Düzeltme isteği - 1 Mayıs 2007). Accepted for publication - August 23, 2007 (Yayın için kabul tarihi - 23 Ağustos 2007).*

◆ *Correspondence (İletişim adresi): Dr. Çağlar Çallı. İzmir Atatürk Eğitim ve Araştırma Hastanesi, Kulak Burun Boğaz Hastalıkları Kliniği, Basın Sitesi 35360 İzmir, Turkey. Tel: +90 232 - 244 72 90 Fax (Faks): +90 232 - 339 00 02 e-mail (e-posta): caglarcalli@hotmail.com*

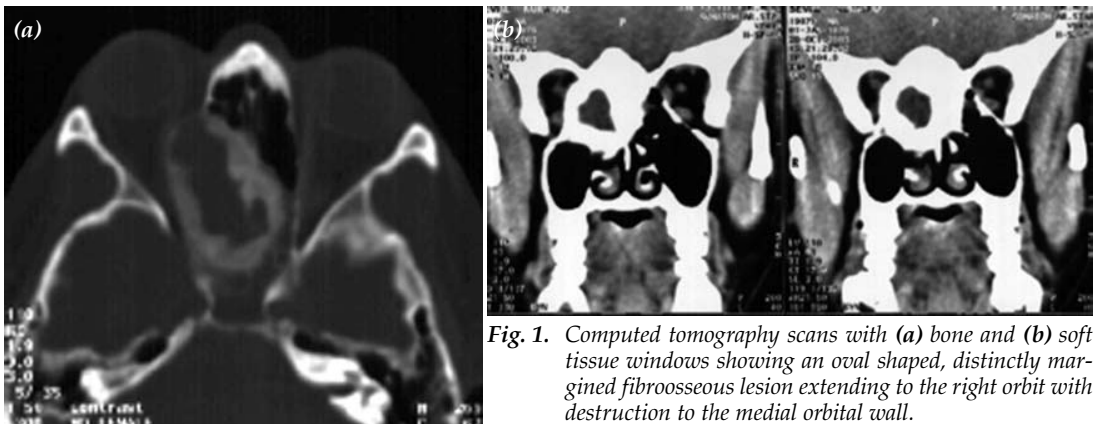

Fig. 1. *Computed tomography scans with* ***(a)*** *bone and* ***(b)*** *soft tissue windows showing an oval shaped, distinctly margined fibroosseous lesion extending to the right orbit with destruction to the medial orbital wall.*

CASE REPORT

A 25-year-old woman presented to the hospital with a history of exophthalmos, headache, and nasal congestion of six-month history. Physical examination showed a well-delineated firm mass on the right side of the nasal septum and right-sided exophthalmos. Eye movements, vision, and the fundus were found to be normal. Axial and coronal computed tomography scans showed a mass in the right ethmoid sinus, extending from the right orbital rim to the right nasal cavity. The mass was well-delineated, round in shape, 4x4.5x3 cm in size, and had central radiopacity and a peripheral dense rim (Fig. 1). The lesion abutted the sphenoidal sinuses mostly on the right side posteriorly and showed mucosal polypoid proliferation and obstructions in the bilateral sphenoidal-right maxillary and frontal sinuses. Multiplanar views of magnetic resonance imaging showed more details (Fig. 2). Radiological findings of the mass were consistent with a benign lesion, most probably a fibroosseous lesion. To relieve the obstruction and the mass effect of the lesion, near-total excision of the mass was performed by a lateral rhinotomy and medial maxillotomy approach. Tissue pathology showed woven bone often rimmed by osteoblasts lying in the layers of lamellar bone (Fig 3). There were woven areas of irregular shape and with peripheral lamellar maturation. Fibrous dysplasia and ossifying fibroma were considered in the differential diagnosis. Based on histologic and radiological findings, the diagnosis was made as ossifying fibroma.

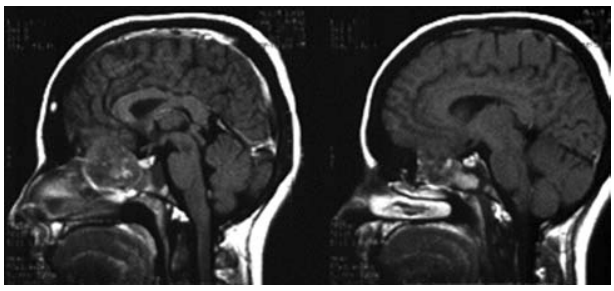

Fig. 2. *Sagittal post-contrast T_1-weighted magnetic resonance images showing peripheral fibroosseous part.*

DISCUSSION

Fibroosseous lesions in the face and jaw are cemento-ossifying dysplasia, fibrous dysplasia, and cemento-ossifying fibroma. Radiological imaging is the key to the diagnosis since all FOLs have a similar pathologic appearance.[5] Cemento-ossifying fibroma originates from mesenchymal blast cells. It is a rare, expansile, benign tumor that predominantly involves the maxillary (approximately 10–20% of cases) and mandibular (approximately 75%) bone. In rare cases, the tumor may involve the nasal cavity and long bones. Patients with a COF usually present within the second to fourth decades of life, with a

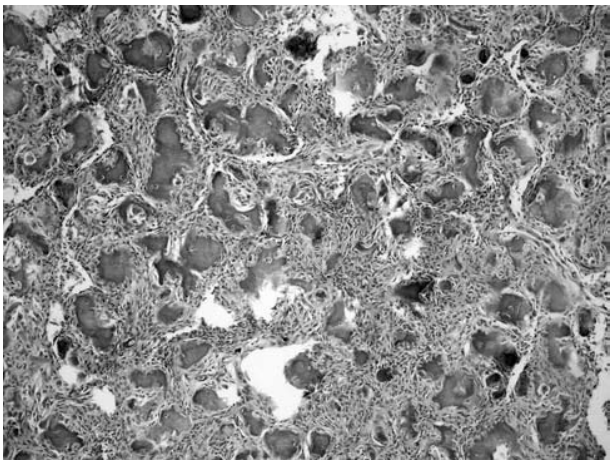

Fig. 3. *Histopathologic view showing osteoblastic proliferation in fibroblastic stroma (H-E x 250).*

higher frequency of females.[4-6] On histopathologic evaluation, COF exhibits lamellar mature bone and closely packed spindle cells. In fibrous dysplasia, the stroma is more collagenized and less cellular, with abortive bony trabeculae and woven trabecular bone. However, similar bone trabeculae may be found in ossifying fibroma, as well.[4-6]

Ossifying fibroma and cemento-ossifying fibroma are now considered to be the two extremes of the same spectrum, because both frequently contain bone and cementum-like tissue.[5,6] These lesions are now called cemento-ossifying fibroma; in contrast to FOL, they are well-defined, and round or oval in shape.[5,6] They are mostly radiolucent, have central radiopacity, and are completely opaque. The mandibula and maxilla are the most affected sites, but rarely the ethmoid sinus, orbits, and the nasal fossa may be involved.[3,4] It has also been reported in auricular soft tissue.[7-10]

Histologically, cementum represents mineralized dental tissue surrounding the tooth apex. The ectopic tissue may cause atypical COF in the ethmoidal region.[5,6] Clinical findings vary depending on the site of involvement and the size of the lesion. In this type of tumors, clinical diagnosis can be easily established especially by computed tomography.[4-7] Magnetic resonance imaging also provides valuable information about the extent and characteristics of the lesion. Fibroosseous lesions usually present as a low-to-intermediate signal intensity on T_1-weighted images and variable signal intensity on T_2-weighted images. After contrast administration, the thick outer layer of the lesion shows intense enhancement. In our case, the lesion showed peripheral enhancement, differing from that seen in fibrous dysplasia. Fibrous dysplasia exhibits low signal intensity on T_2-weighted images, whereas ossifying fibroma may have high signal intensity, as seen in our case. In fibrous dysplasia, contrast enhancement involves only the expanded bone; however, in ossifying fibroma, enhancement occurs in the outer shell and septa, as in our case. These findings help in the differentiation of ossifying fibroma from fibrous dysplasia.

Prognosis is good after surgical excision. Mandibular lesions usually grow slowly, but lesions in the ethmoid sinus are locally aggressive and may cause nasal obstruction, sinusitis, headache, proptosis, diplopia, rhinorrhea, and may extend to the anterior cranial fossa.[3,5,6] In our patient, there was destruction of the medial wall of the orbit and orbital extension.

Computed tomography is the most accurate method for showing calcifications and destruction in the advanced stage of this type of tumors.[3-7] This was the case in our patient. Osteosarcomas, chondrosarcomas, epithelial malignant tumors should also be included in the differential diagnosis of tumoral lesions in the ethmoid sinus. An aneurysmal bone cyst accompanying cemento-ossifying fibroma was also reported in the mandibular corpus.[8] Although all have different clinical, pathological and histological findings,[4,5] imaging is the key factor for diagnosis and follow-up of FOLs.

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