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Case report

Myxoma of the Zygomatic Bone – A Case Report

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A B S T R A C T

Myxoma is a benign tumor composed of primitive connective tissue cells and mesenchymal mucousal stroma. Also referred to as, a gelationus or colloidal tumor. Although rare, it can be found in the atrium of the heart, and it is the most common heart tumor. It has also been described in other body sites, one of which is the bone. We report a case of a 57-year-old female patient, with recurrent headaches located in the area of the right half of the face. Radiological analysis (Multislice Computed Tomography of the paranasal sinuses and viscerocranium) was performed, and a formation of irregular contours, destroying the right zygomatic bone, was described, measuring 25x17x20 mm in its widest diameters. Its medial border was adjacent to the lateral wall of the right maxillary sinus and the cortical bone in this segment was thinned, but preserved. A probatory excision was performed in general anesthesia, and the histopathological finding showed, star-like tumor cells embedded in mucoid stroma and infiltrating the bone. After pathohistological confirmation of myxoma, the tumor was excised in total, using infraorbital surgical approach to the zygomatic bone. During the follow-up, the patient was symptom free, without headaches, and there were no signs of local tumor recurrence. Despite of the fact that myxoma behaves as a benign disease in its nature, it can cause destruction of the tissue in the vicinity of the tumor itself, and thus major health issues for the patient. A timely proper diagnosis and the right choice of a surgical treatment can help avoid more extensive surgery procedures, as shown in our case report.

Key words: *myxoma, zygomatic bone, bone destruction, surgical aproach*

Introduction

Myxoma is a benign tumor composed of primitive connective tissue cells and mucousal stroma. Also referred to, as gelatinous or colloidal tumor.

Although rare, it can be found in the atrium of the heart and it is the most common heart tumor. It can develop in a variety of locations, including subcutaneus and aponeurotic tissues, bones, genitourinary tract, skin, retroperitoneum, intestine, pharynx, joints and sceletal muscles[1].

Myxoma rarely occurs[2] in the head and neck region, and in this region it is usually related to odontogenic myxoma of the maxilla and the mandibula, making 3–6% of all odontogenic tumors[3].

Case Report

We report a case of a 57 years old female patient, with recurrent headaches located in the area of the right half of the face.

The patient had no remarkable medical history. Except for headeches, she did not report other symptoms. She denied previous head trauma of immunodeficiency symptoms and family history was negative for similar disorders.

Radiological analysis (Multislice Computed Tomography of the paranasal sinuses and viscerocranium) was performed, and a formation of irregular contours, destroying the right zygomatic bone, was described, measuring 25x17x20 mm in its widest diameters. Its medial border was adjacent to the lateral wall of the right maxillary sinus and the cortical bone in this segment was thinned, but preserved (Figure 1). Considering the MSCT (Multislice Computed Tomography) findings, in an initial surgical procedure the right maxillary sinus was explored endoscopically, but the tumorous mass was not found. An external infraorbital probatory excision followed, based upon the radioglogist s MSCT findings and descrption.

The histopathological finding showed, star-like tumor cells embedded in mucoid stroma and infiltrating the bone (Figure 2). After pathohistological confirmation of myxoma, the tumor was excised in total, using infraorbital surgical approach to the zygomatic bone (Figure 3a and b).

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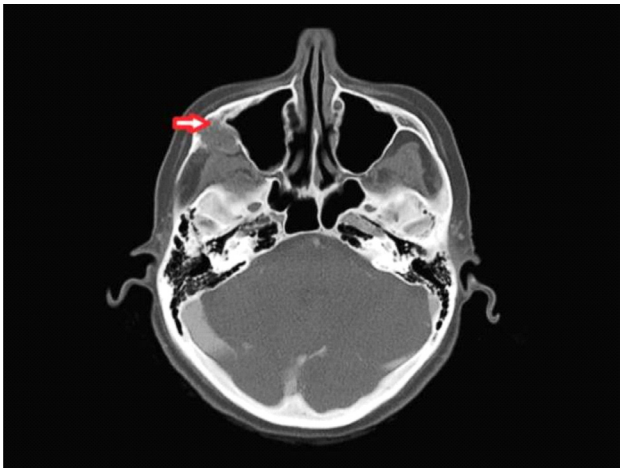

Fig. 1. Radiological analysis (Multislice Computed Tomography of the paranasal sinuses and viscerocranium) showing a formation of irregular contours, destroying the right zygomatic bone, measuring 25x17x20 mm in its widest diameters.

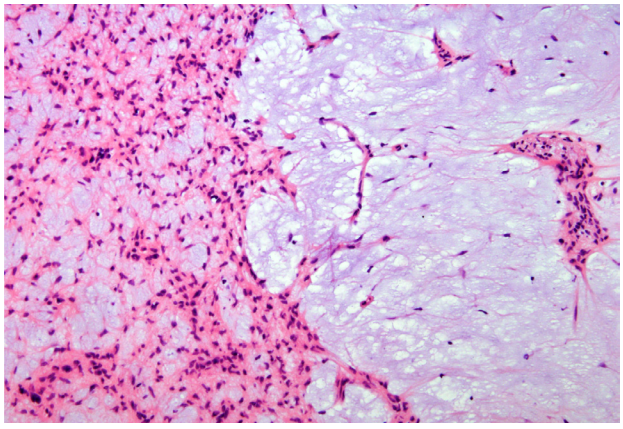

Fig. 2. The histopathological finding: star-like tumor cells embedded in mucoid stroma (hemalaun-eosin staining, 40x).

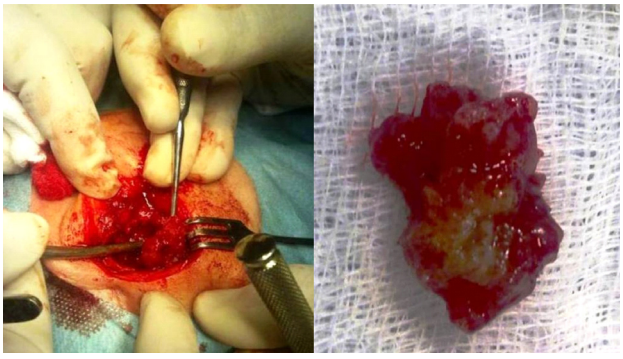

Fig. 3. a) Infraorbital surgical approach to the zygomatic bone. b) Tumor tissue.

During the follow-up, the patient was symptom free, without headaches, and there were no signs of local tumor recurrence.

At check up, a 18F-fluorodeooxyglucose Positron Emission Tomography/Computed Tomography (18F-FDG PET/CT) was performed and no abnormal findings were found.

Discussion and Conclusion

Myxoma is the most common primary cardiac tumor and is classified as benign in nature[4]. Suspicion for this diagnosis is raised in patients with cardiac syptoms or if throughout cardiac work-up, a tumor mass is found in cardiac cavities, particularly in the left atrium.

The very first case of myxoma was described by Virhow in 1871[5,6], but diagnostic criteria and myxoma definition was suggested by Stout in 1848[6,7].

This tumor incidence in the heart muscle is aproximately 0.5 per one million inhabitants per year[8] and intramuscular myxoma have an approximate incidence of 1 per million of the population per year[9]. Incidence of myxoma tumor in bone structures is lower than mentioned above, and it is usually related to odontogenic tumors of the maxilla and the mandibula, making 3–6% of odontogenic tumors overall[10]. The etiology for myxoma has been unknown to this day. Familiar variants exist[11], but most cases are sporadic, and more often found in middle aged female population[12], as is was in our case, of a 57 years old female patient with unrelated familial history.

For some authors, chronic inflammation has the leading role in the etiology of myxoma[13], others relate it to immunosuppression, for example after organ transplantation[13]. A third group of authors describe hypoxia as a trigger, creating appropriate microenvironment for tumor existance, addditionally activating vascular endothelial growth factor (VEGF) A, as an angiogenesis and vascular permeability promotor[14].

Cases of myxoma related to chromosome 12 and 17 abnormalities have been described in literature[15]. Often is the myxoma an accidental finding in symptom free patients.

In our patient' s case, the main struggle was a pulsating headache in the right half of the face, in the projection of the right maxillary sinus.

It is important to accentuate the clinical significance of bone lesions as showed in our case report.

In differential diagnosis, enteties that should be considered are: aggressive angiomyxoma, myxoid neurofibroma, myxoid liposarcoma, low-grade myxofibrosarcoma, cellular myxoma, juxta-articular myxoma and nodular fasciitis[17]. Usually, when myxoma becomes symptomatic in other locations than the heart, such as bone lesions, as it was the case with our patient, seldom is myxoma the first diagnosis clinicians think of. After pathohistological confirmation of myxoma, the tumor was excised in total, using infraorbital surgical approach to the zygomatic bone. Because of the existance of Mazabraud's syndrome described in literatue, characterized by the association of single or multiple intramuscular myxomas with fibrous

dysplasia which can develop in a single bone or in multiple bones[16], we performed 18F-fluorodeooxyglucose Positron Emission Tomography /Computed Tomography (18F-FDG PET / CT). No bone lesions or fibrous dysplasia were found.

This myxoma case is also interesting for its unusual location. In the attainable literature, only three cases of zygomatic bone myxoma have been described so far.

Regardless of a non-metastatic tumor behavior, myxoma is locally invasive in nature and characterised by slow, often, non-symptomatic growth, sometimes resulting in expansion and even perforation of the cortex of the involved bone[18].

Fortunately, due to exact diagnosis and prompt and efficient surgical treatement, progression and bone destruction was succesfully prevented in our patient's case.

The patient from this case report is currently asymptomatic, showing no signs of tumor recurrence. She is coming for regular check-ups, because of myxoma's tendence to recidivation, often described in medical literature[19].

In conclusion, despite the fact that myxoma behaves as a benign disease, it can cause destruction of surrounding tissues and structures, thus imparing the patient's well being and life quality. Early diagnosis and surgical treatment can help avoid more extensive surgical procedures, as shown in our case report.

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MIKSOM ZIGOMATIČNE KOSTI – PRIKAZ SLUČAJA

SAŽETAK

Miksom je dobroćudni tumor građen od primitivnih stanica vezivnog tkiva i mezenhimne sluzave strome. Naziva se još i gelatinoznim ili koloidnim tumorom. Iako rijedak, može se naći u srčanim pretklijetkama i najčešći je tumor srca. Opisan je i na drugim lokalizacijama, a jedna od njih je i kost. Prikazujemo slučaj 57-ogodišnje pacijentice koja se javlja zbog recidivirajuće glavobolje locirane u području desne polovice lica. Učinjenom radiološkom obradom (Višeslojna kompjuterizirana tomografija paranazalnih sinusa i viscerokranija) uoči se formacija nepravilnih kontura koja destruira desnu zigomatičnu kost. U najširim promjerima opisana formacija je veličine 25x17x20 mm. Svojim medijalnim rubom naslanja se uz lateralnu stijenku desnog maksilarnog sinusa, kortikalna kost u tom segmentu je stanjena, ali održana. U općoj anesteziji učini se probatorna ekscizija te patohistološki nalaz pokaže zvjezdolike tumorske stanice koje infiltrativno rastu prema kosti uklopljene u mikoidnu stromu. Postavi se dijagnoza miksoma zigomatične kosti koji se u daljnjem postupku u potpunosti ekscidira infraorbitalnim pristupom na zigomatičnu kost. Tijekom perioda praćenja i kontrolnih pregleda,pacijentica je asimptomatična, bez znakova recidiva. Unatoč tome što se miksom predstavlja kao dobroćudna bolest, može biti uzrokom destrukcije tkivnih struktura u okolini tumora, a time i tegoba bolesnika. Pravodobna dijagnoza te operacijsko liječenje mogu izbjeći opsežnije operativne zahvate, kao što je bilo u prikazanom slučaju.