

Synovial sarcoma of temporomandibular joint -A case report

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Abstract

Introduction: Synovial sarcoma (SS) is a rare highly malignant soft tissue tumor of young adults. It generally develops in close proximity to joints of extremities but contrary to its name, it does not originate from synovial cells but is of mesenchymal origin. Upto 10% of soft tissue sarcomas are synovial sarcomas. SS of the Temporomandibular joint (TMJ) is a very rare entity. Here, we report a case of synovial sarcoma of the temporomandibular joint in a 22 year old male patient. Case report: A Male patient, 22 years of age had a complaint of a painful swelling in the left pre-auricular region since 2 months. CBCT of the left TMJ revealed advanced Osteolytic changes in the left condylar head region. The affected region had an irregular 'moth-eaten appearance'. USG guided FNAC revealed biphasic pattern of malignant cells with both epithelial and stromal elements. Histopathology of the tumor showed a cellular tumor with biphasic appearance. Immunohistochemistry (IHC) results showed positivity for CK (MNF) which is expressed in epithelial elements. 70% of the spindle cells expressed TLE-1 positivity and moderate expression for BCL-2 and CD 99. The histomorphological and IHC findings were consistent with the diagnosis of biphasic synovial sarcoma.

Conclusion: Synovial Sarcomas are very rare in the TMJ region. A CBCT investigation can best define the outline of such tumors, degree of bone involvement & destruction. A high index of suspicion along with CBCT and radiological evaluation, FNAC and histopathological assessment usually clinches the diagnosis.

Keywords: Synovial Sarcoma, TMJ, CBCT, Synovium.

Introduction

Synovial sarcoma (SS) is a rare highly malignant soft tissue tumor of young adults. It generally develops in close proximity to joints of extremities but contrary to its name, it does not originate from synovial cells but is of mesenchymal origin. Upto 10% of soft tissue sarcomas are synovial sarcomas.¹ It is a very rare tumor and according to a study done in Taiwan, the annual incidence of SS is approximately 0.08 per 100000 person years.² The incidence in the United States is around 800 cases every year.³ It typically occurs in adolescents and young adults with a mean age of 36 years out of which about one third cases are below 20 years of age.³ There is no sex predilection as men and women seem to be affected almost equally. SS commonly affects the joints of the extremities but SS of the Temporomandibular joint (TMJ) is a very rare entity.

Here, we report a case of synovial sarcoma of the temporomandibular joint in a 22 year old male patient.

Case Report

A male patient, 22 years of age had a complaint of a painful swelling in the left pre-auricular region since 2 months. The swelling was associated with frequent headaches on left side, pain in the ear & restricted jaw movements. There was a localized swelling measuring 2.5× 3.5cm in size in front of the left ear. The swelling was tender, firm in consistency & non-compressible.

There were no significant changes noted in the oral cavity. Two dimensional radiograph of the Joint did not reveal any significant information.

CBCT of the left TMJ revealed advanced Osteolytic changes in the left condylar head region. The affected region had an irregular 'moth-eaten appearance'. There was periosteal activity & ragged borders at the periphery of the lesion. The inter-articular joint space had increased to 17mm indicating an aggressive neoplastic lesion, the most common at that age being TMJ Sarcoma.

His routine blood investigations were within normal limits. A Mantoux test done to rule out a tubercular etiology was also negative.

USG guided FNAC revealed biphasic pattern of malignant cells with both epithelial and stromal elements. The epithelial component consisted of malignant cells with high N:C ratio while the stromal component consisted of malignant spindle cells with round to ovoid plump nuclei. A provisional diagnosis of a sarcoma was made. A left hemimandibulectomy from TMJ upto the angle of mandible was performed. Histopathology of the tumor showed a cellular tumor with biphasic appearance consisting of fascicles of short spindled cells and fewer areas showing cleft like glandular and nested pattern of cells with epithelial morphology. There was focal mild to moderate atypia with a mitotic count of 6/10 hpf.

A small area of necrosis occupying much less than 50% of the tissue was also noticed with sprinkling of mast cells and destruction of the bone. There was no

lymphovascular invasion. FNCLCC score =histology score(3)+mitoses score(1)+necrosis score(1)=5=Grade 2 (on a scale of grade 1 to3)was made. The mandible and the zygomatic arch were free of tumor. Immunohistochemistry (IHC) results showed positivity for CK(MNF)which is expressed in epithelial elements. EMAs weakly expressed in a small focus of epithelial elements. The spindle cell element was negative for both CK and EMA. 70%of the spindle cells expressed TLE-1 positivity and moderate expression for BCL-2 and CD 99. S100, SMA, Desmin and CD 34 were all negative. The histomorphological and IHC findings were consistent with the diagnosis of biphasic synovial sarcoma.



Fig. 1: OPG (2D) image revealing no significant finding in the left TMJ region



Fig. 2: Clinical image of the swelling on the affected side

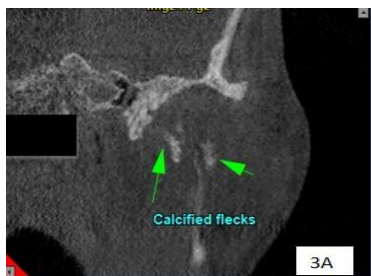


Fig. 3A



Fig. 3B



Fig. 3C

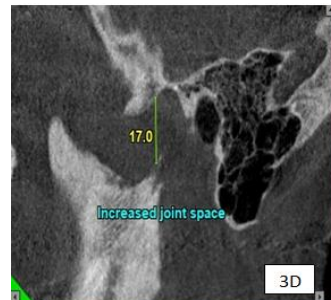


Fig. 3D

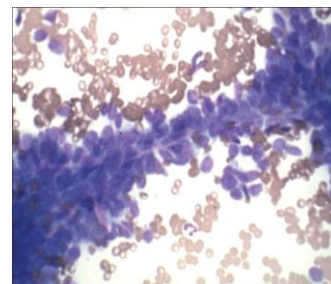


Fig. 4: FNAC 40x image showing sheets of malignant epithelial cells

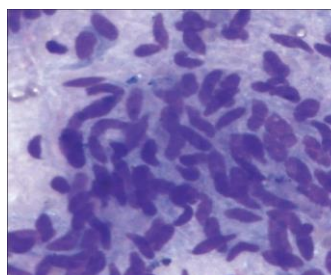


Fig. 5: FNAC image 40 x showing spindle cells

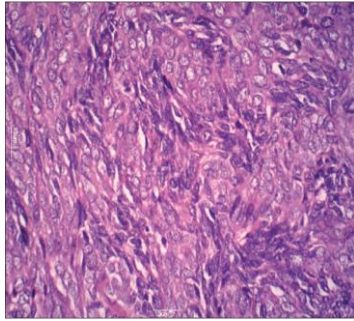


Fig. 5: H&E stain of 40x

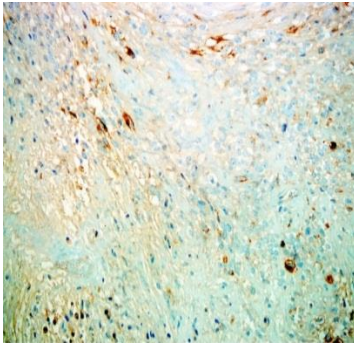


Fig. 6: IHC 40X EMA positive epithelial cells

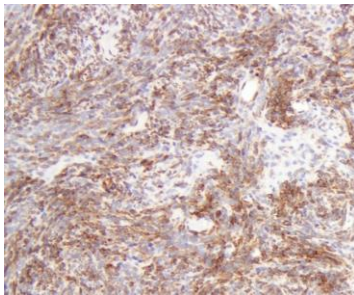


Fig. 7: 10x BCL 2 positive cells

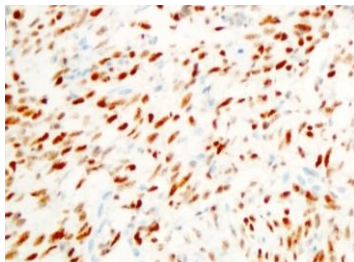


Fig. 8: Showing TLE 1 positive cells

Discussion

Synovial sarcomas are rare soft tissue neoplasms which arise from primitive mesenchymal tissue. Simon reported the first case of SS way back in 1865.⁴ However, Lejars and Rubens –Duval first gave a detailed description of SS in 1910.⁵ The term “synovial sarcoma” was coined by Haagensen and Stout in 1944, who described it as a distinct clinical and histological entity.⁶

SS is a rare soft tissue neoplasm but does not arise from synovial region of the joints, tendons and bursae but appears to arise *denovo* from primitive mesenchymal cells. SS of the head and neck do not have a tendency to metastasize but 20% cases may have regional lymph node metastasis.^{7,8} It has a propensity to spread by direct extension along the tissue planes and also by vascular route, hence the resemblance to sarcoma. Although a few cases have been diagnosed on 2D radiographs, this is the first case diagnosed on CBCT imaging.

Typically, SS are oval, locular masses with a false capsule around it. Cut surface shows a smooth whirling pattern and may have solid and cystic components with areas of haemorrhage and necrosis. Our case had a solid and a major cystic component. Histologically, SS may be biphasic with both epithelial and spindle cell component or may be monophasic with only a spindle cell component. Biphasic SS is more common than the monophasic variant. Rarely, a poorly differentiated SS may be seen which has a very poor prognosis.

Treatment consists of complete surgical removal of the tumor tissue locally before metastasis has occurred. Role of radiotherapy is controversial with some authors favouring RT while others are of the opinion that it should be used as an adjunct to surgical removal. However, Krugmen et al observed a more favourable outcome when RT was used post operatively.⁹

Chemotherapy has a very limited role. Prognosis of the tumor depends upon the time at diagnosis, extent of the tumor and metastasis. Females have a better prognosis than males. Survival rates may vary from 29-50% but head and neck tumors having a better prognosis.¹⁰⁻¹²

Nomura et al described four cases of SS of TMJ.¹³ They concluded that local control of tumor has improved but death due to distant metastasis is still a major issue. Luo et al reported a case of biphasic SS of TMJ with symptoms of pain, tinnitus and hearing loss.¹⁴ Stadelmann W.K. et al described SS and its treatment modalities while reporting their case of SS of TMJ.¹⁵ N. Younus et al reported a case of biphasic SS of head and neck region in a 30 year old Saudi woman arising from the posterior limit of the thyroid cartilage.¹⁶ Immuno histochemically, TLE 1 is a sensitive and specific marker for SS.¹⁷ The spindle cells have strong expression for vimentin and moderate expression of Cytokeratin especially in the biphasic variant of SS. Other markers such as CK7, CK19 and EMA are also positive. As was demonstrated in our case too. Cytogenetically more than 90% of SS demonstrate $t(x;18)(p11.2;q11.2)$ translocation of chromosomes and is diagnostic of SS.¹⁸⁻²⁰

Conclusion

Synovial Sarcomas are Malignant tumors of the soft tissue. They are very rare in the TMJ region (5% to 6%). They may be easily misdiagnosed as they produce common signs and symptoms like swelling, decreased

jaw movements, TMJ pain, Headaches, malocclusion, etc. A CBCT investigation can best define the outline of such tumors, degree of bone involvement & destruction. It has an added advantage of using minimal radiation and being non-invasive. CBCT can easily capture early tumors leading to good prognosis. A high index of suspicion along with CBCT and radiological evaluation, FNAC and histopathological assessment usually clinches the diagnosis.

Conflict of Interest: None

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