Tenosynovial giant cell tumor of temporomandibular joint – A case report with literature review

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Introduction

Tenosynovial giant cell tumor (TGCT) is a rare tumor, arising in the synovial lining of joints, tendon sheaths and bursae. It more commonly involves digit or large joints. TGCT involving temporomandibular joint (TMJ) is rare with only more than 100 cases being reported as of 2021 [7]. We report a case of tenosynovial giant cell tumor involving temporomandibular joint in a 47-year-old female with a brief literature review.

Clinical history

The patient was a 47-year-old Chinese female who presented with blocked ear sensation on the left side for 3 years. MRI and CT scan showed a soft tissue mass measuring 3.5 cm x 5.3 cm x 5 cm extending to left temporal and parietal lobes superiorly, abutting left cavernous sinus medially, left buccinator and left temporalis muscle laterally and extending to left temporomandibular joint inferiorly (Figure. 1). Biopsy was performed and later followed by radical excision of the lesion.

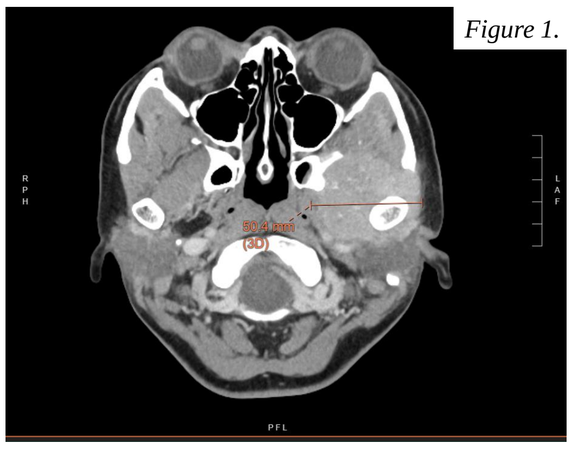

Figure 1.

Surgical findings and pathological features

During operation, a brownish hard tumor pushing the dura of temporal base was identified. Dura was intact. The tumor showed encasement of mandibular condyle.

Fragments of brownish tumor tissue attaching to the temporomandibular joint were received. Part of mandibular condyle was included. Specimen measured 4 x 3 x 2.5 cm. (Figure. 2)

Figure 2.

Microscopic examination

Sections showed the tumor tissue, which attached to synovium (Figure. 3), composed of mixture of mononuclear cells and multinucleated osteoclast-like giant cells. Tumor cells were arranged in diffuse sheets (Figure. 4). Foamy macrophages were occasionally seen. Chondroid metaplasia was noted focally (Figure. 5). The mononuclear cells possessed distinct cytoplasmic border, moderate amount of eosinophilic cytoplasm, oval nuclei and occasional intracytoplasmic hemosiderin deposit (Figure. 6). High grade atypia and necrosis were not seen. Mitotic activity was occasionally noted.

Immunohistochemically, mononuclear cells were positive for D2-40 and some of them also expressed clusterin. Both mononuclear cells and multinucleated giant cells were positive for CD68. They were negative for H3.3 G34W and H3.3 K36M immunostains. (Figure. 7)

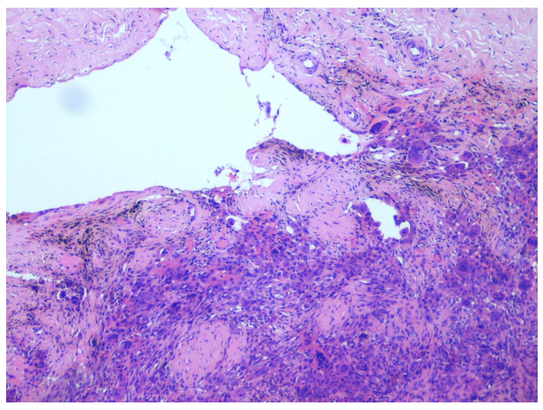

Figure 3. Tumor attached to the synovium

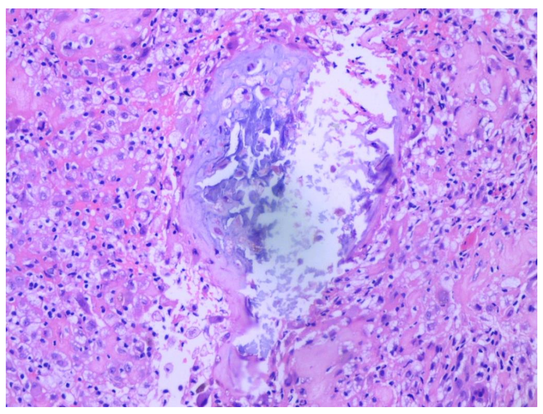

Figure 5. Focus with chondroid metaplasia

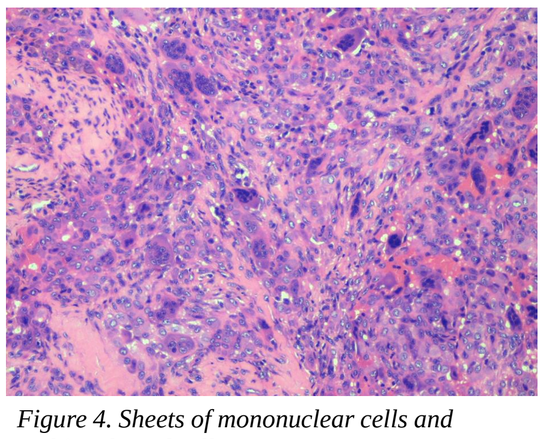

Figure 4. Sheets of mononuclear cells and multinucleated cells

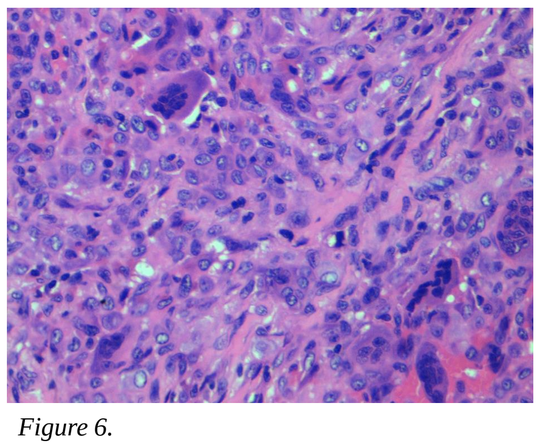

Figure 6.

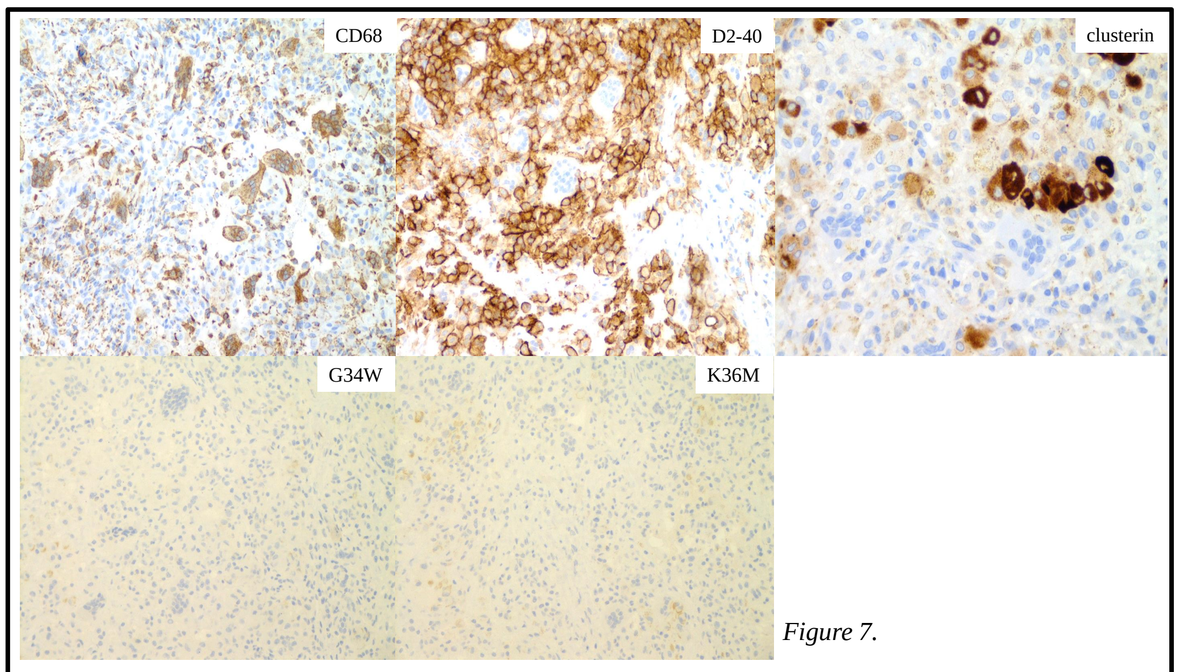

Figure 7.

Progress

No evidence of recurrence was identified 2 months after operation.

Discussion

TGCT is a rare tumor, arising in the synovial lining of joints, tendon sheaths and bursae. It is classified as localized and diffuse type. Translocation involving the CSF1 gene (colony stimulating factor 1) and overexpression of the CSF1 protein are demonstrated in some TGCT [1]. It usually occurs in patient aged from 30-50 years. The incidence rate of TGCT is estimated 29, 10, and 4 per million person-years for digit, localized-extremity and diffuse TGCT respectively [6].

TGCT involving TMJ is even rarer with more than 100 cases being reported as of 2021 [7]. Majority of tumors affecting the TMJ are extraarticular, locally destructive and often extend to the temporal bone [10]. Clinical presentation varies and are non-specific. Patient can present with pain in TMJ, trismus, ear fullness, hearing loss, tinnitus and preauricular swelling [1-5,7-10].

Imaging studies including MRI and CT serve as the major modalities for investigation. Lesion manifests as an expansile soft tissue mass with encasement of mandibular condyle and displacement of dura of middle cranial fossa. 'Blooming effect' on MRI study has been described characteristic for TGCT due to the presence of hemosiderin in the lesion [1, 3]. Overall features of TGCT are non-specific and there is usually a delay in diagnosis. Preoperative duration ranges from 6 months to 180 months[1].

Microscopic examination reveals diffuse sheets of mononuclear cells, multinucleated giant cells, histiocytoid cells with hemosiderin deposition [1-8]. Bone destruction and dystrophic calcification are also seen. Dystrophic calcification may mimic chicken-wire pattern of calcification in chondroblastoma. Chondroid metaplasia has been reported in TGCT involving TMJ and it is a predilection for the lesions in TMJ [1,3]. Immunohistochemical study shows mononuclear cells are positive for CD68, D2-40 and clusterin. S100 expression is found in area with chondroid metaplasia [1,4].

Surgical treatment, wide local excision, is the mainstay of treatment. Curettage and limited resection carry a higher rate of recurrence. A limited cohort study reports 15-29% rate of recurrence. Frequent imaging follow up is advised for subtotal resection [3]. Adjuvant radiotherapy is an option to patient with recurrent disease [5]. Other adjuvant treatments including cryosurgery and immunotherapy (tumor necrosis factor (TNF)-alpha blocker and CSF-1 receptor directed targeted therapy) are also cited [1]. TGCT involving TMJ is usually a locally destructive disease, yet rare malignant form of TGCT with metastatic lesion in lungs and lymph nodes has been documented [10].

Conclusion

TGCT involving TMJ is a rare disease with non-specific clinical and radiological features which lead to a delay in diagnosis. Completeness of surgical excision is related to the rate of recurrence and wide local excision is considered curative.

Reference

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