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Diffuse-type Tenosynovial Giant Cell Tumor of the Temporomandibular Joint with Intracranial Extension

Purpose:

Tenosynovial giant cell tumors occur in two types – a more indolent localized form (nodular tenosynovitis) which primarily occurs in hands and feet, and the more infiltrative diffuse-type Giant cell tumor (dt-GCT)[2]. Dt-GCT can occur in any synovial joint space, most commonly in the knee; it may also be found less commonly in the hips, ankle and elbow, and in rare cases in the temporomandibular joint (TMJ)[1]. This presentation will describe a rare dt-GCT of the TMJ, its imaging appearance, and its subsequent total resection and reconstruction.

Description:

A 36-year-old gentleman presented with a five-year history of a lump on the left side of his head. Although initially asymptomatic, he now experienced difficulty opening his jaw, clicking, headaches, tinnitus, and numbness in the left V2 distribution. CT demonstrated a 4.3 cm lytic, predominantly hyperdense and heterogeneously enhancing lesion involving the left temporal bone with a soft tissue component, also extending to the left sphenoid bone, the left infratemporal region, and the left temporomandibular joint. On MRI, the lesion was T1 hypointense, T2 predominantly hypointense with internal T2 hyperintensities. On post-contrast imaging, the lesion was primarily hypoenhancing with internal septations and peripheral rim enhancement. The mass also extended inferiorly into the masticator space, involving the lateral pterygoid muscles and the temporomandibular joint. There was intracranial extension with mass effect on the left temporal lobe and asymmetric dural enhancement. The patient underwent gross total resection with final pathology demonstrating diffuse-type tenosynovial giant cell tumor with extensive bone involvement.

Summary:

There are two types of tenosynovial giant cell tumors – a more localized form involving tendons (nodular synovitis) and a diffuse type which typically has a monoarticular origin[2]. This patient follows a typical clinical course of TMJ dt-GCT with a growing mass presenting with progressive pain, tinnitus, clicking, difficulty opening jaw, and sensory disturbance. The imaging appearance of tenosynovial giant cell tumors is due to iron deposition[1], with hyperdensity on CT and primarily hypointense T1 and T2 signal on MRI with blooming on gradient echo sequences. Surgical resection is the standard treatment of dt-GCT[3], with radiotherapy typically reserved for unresectable tumors or tumors where complete resection was not possible[2]. Long term follow up is recommended over the course of 5-7 years with CT or MRI as 8% of cases reported recurrence of the tumor[2,3].

Citations:

1. van der Heijden L, et al. The management of diffuse-type giant cell tumour (pigmented villonodular synovitis) and giant cell tumour of tendon sheath (nodular tenosynovitis). *J Bone Joint Surg Br*. 2012 Jul;94(7):882-8. doi: 10.1302/0301-620X.94B7.28927.
2. Bredell M, Schucknecht B, Bode-Lesniewska B. Tenosynovial, diffuse type giant cell tumor of the temporomandibular joint, diagnosis and management of a rare tumor. *J Clin Med Res*. 2015;7(4):262–266. doi:10.14740/jocmr1872w
3. Romañach MJ, Brasileiro BF, León JE, Alves DB, de Almeida OP, Vargas PA. Pigmented villonodular synovitis of the temporomandibular joint: case report and review of the literature. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod*. 2011 Mar;111(3):e17-28. doi: 10.1016/j.tripleo.2010.11.019.

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