CASE REPORT

Giant Solitary Synovial Chondromatosis of the Temporomandibular Joint with Intracranial Extension

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Abstract—Synovial chondromatosis are rare entities but are well-described lesions in the literature that can affect many joint areas of the body. A case of tumoral synovial chondromatosis involving the temporomandibular joint with intracranial extension through mandibular fossa is reported. As long as there was significant infratemporal and extradural invasion of the middle and posterior fossa, a transtemporal and infratemporal approach was performed and total removal of the lesions was achieved. A brief review of skull base synovial chondromatosis is presented.

Synovial chondromatosis is an idiopathic metaplasia of the synovial cells to cartilage, which often calcifies and may form intra-articularly and/or extra-articularly. Secondary synovial chondromatosis may be polyarticular and is associated with slowly progressive degenerative joint disease, trauma and neurological disease. Primary synovial chondromatosis is monoarticular, twice as common in men as in women over 60 years of age. Sites of involvement include the elbows, shoulder, knee, hip, and ankles.¹

The chondroma of a giant solitary synovial chondromatosis is larger and measures from 1 cm to over 20 cm.² They may occur in any joint, including the temporomandibular joint, formed by the fusion of multiple synovial chondromas or by continued growth of a solitary synovial chondroma into a large, well-circumscribed mass. Radiographically, they may be difficult to distinguish from chondrosarcoma^{3,4} and histologically they may have a similar appearance to low-grade chondrosarcoma.⁵

The following case presents a patient with a giant solitary synovial chondromatosis of the temporomandibular joint and only one case was published to our knowledge in the literature with intracranial extension.⁶

MATERIALS AND METHODS

Case Report

A 65-year-old female had progressive neurological symptoms that began with pain of the left side of the face 8 years ago. Approximately 4 years after the onset of these symptoms, she developed weakness and sensory disturbance of the left side of the face, difficulty in swallowing, hoarseness, and growing of a preauricular tumor.

At admission in our department, a neurological examination revealed that the patient had left-sided peripheral facial nerve palsies, hearing impairment, weakness of the left soft palate, atrophy of the left tongue, and a protrusion at the left preauricular zone toward frontal scalp, 8 cm in diameter, hard, tender, smooth-surfaced, and covered by normal skin.

CT scan revealed a 5 × 6 cm destructive mass occupying the middle fossa with extension into the petrous bone, posterior fossa, and temporomandibular joint. Radiograph showed erosion of the mandible and condylar fossa (Figs. 1 and 2). MRI localized an enhancing lesion

Figure 1. CT scan revealed a 5 × 6 cm destructive mass occupying the middle fossa with extension into the petrous bone, posterior fossa, and temporomandibular joint.

centered in the middle fossa with extension into the petrous bone and toward the temporal lobe extradurally (Figs. 3 and 4) and inferiorly, into the temporomandibular joint and infratemporal space (Figs. 5 and 6). Arteriography demonstrated an avascular lesion without displacing the carotid artery.

Surgical Technique

Surgery was performed with the patient in a semi-lateral position and the head laterally fixed to a Mayfield headrest. The surgical field was prepared including the abdomen, if an abdominal fat graft or a rectus abdominal muscle free flap was needed. A large C-shaped incision, from just superior to the pterion and extended superiorly and posteriorly in a curvilinear fashion to the postauricular area and into the neck. The dissection was performed closing the external auditory meatus and reinforcing it with the aid of a fibroperiosteal flap. Dissection of the cervical skin flap, deep to the platysma muscle, continued anteriorly to the submandibular area and the inferior aspect of the incision was dissected off the lateral and anterior aspect of the sternocleidomastoid muscle. The peripheral main trunk of the facial nerve was identified and dissected from the bony external au-

Figure 2. CT scan revealed a 5 × 6 cm destructive mass occupying the middle fossa with extension into the petrous bone, posterior fossa, and temporomandibular joint.

Figure 3. MRI localized an enhancing lesion centered in the middle fossa with extension into the petrous bone and extradurally toward the temporal lobe.

Figure 4. MRI localized an enhancing lesion centered in the middle fossa with extension into the petrous bone and extradurally toward the temporal lobe.

Figure 6. MRI localized the tumor inferiorly, into the temporomandibular joint and infratemporal space.

Figure 5. MRI localized the tumor inferiorly, into the temporomandibular joint and infratemporal space.

ditary canal to the parotid gland. The periosteum of the zygomatic arch was dissected but it was partially eroded by the tumor and was completely resected. The temporalis muscle was elevated from the temporal fossa and the neck of the condyle of the mandible exposed.

With this exposure the tumor was found to be large and solid white avascular, and seemed to have arisen from the temporomandibular joint, growing through the mandibular fossa to the middle fossa extradurally and eroding skull base from the petrous bone to the pterion. The tumor was easily removed with its capsule from the bone and it was not necessary to perform a temporal craniotomy because the tumor eroded all the zygomatic arch, condyle, mandibular fossa, middle fossa, temporal bone, and partially the petrous bone, growing to the posterior fossa. Dissection of the extradural middle fossa and infratemporal fossa tumor was easily performed (Fig. 7). Reconstruction of the skull base defect with abdominal fat and the temporalis muscle was achieved.

Microscopically, lobulated masses of hyaline cartilage separated by fibrous tissue are visible (Fig. 8). Chondrocytes are within lacunae, which are partially occupied by hyaline material. Foci of calcification are showed. Sinovium is present and metaplasia from fibrous tissue to cartilage could be demonstrated. Chondrocytes show some degree of atypia and focally binucleated forms (Fig. 9, 10).

Figure 7. After debulking the tumor and the bone eroded, dissection of the extradural middle fossa and infratemporal fossa tumor was easily performed.

Figure 8. Low power view showing lobules of cartilage separate concentric layers of collagen (HE, 60 $\times$).

Figure 9. Focal areas of cartilage are better preserved (HE, 100 $\times$).

Figure 10. Lacunae partially occupied for hyaline material and chondrocytes with enlarged nuclei. A binucleated cell is indicated (HE, 400×).

Ten days after surgery the patient noted improvement of swallowing and hoarseness, but there was no significant improvement of the left facial nerve weakness and deafness. A postoperative CT scan showing reconstruction of the defect with fat and the tumor had been completely removed (Fig. 11). Fifteen months after surgery the patient had return of facial function to a House grade III. No tumor recurrence has been evident upon follow-up.

Figure 11. CT scan showing reconstruction of the defect with fat after the tumor had been completely removed.

DISCUSSION

The pathogenesis of synovial chondromatosis is not fully understood but is thought to represent a metaplasia of stromal cells of the synovium to cartilage which often calcifies.¹ To the three phases of synovial chondromatosis currently recognized by Milgram,⁷ other authors add a fourth phase: a large intra- or extra-articular solitary, calcified cartilagenous mass.² However, calcifications are absent in one-third of the patients and occasionally there is bone involvement by pressure erosion. The diagnosis is made mainly on combining the histology with the clinical and radiological features.

The differentiation between synovial chondromatosis and low-grade chondrosarcomas with the clinical and radiological features it is not always possible.⁸ Bertoni et al. defined several histological criteria to distinguish between the nodules of hyaline metaplastic cartilage and the chondrocyte's minimal citological atypia in clusters with abundant intervening matrix in the synovial chondromatosis.⁹

Although approximately 100 cases have been reported in the literature, recently several cases of synovial chondromatosis of the temporo-mandibular joint and their arthroscopic or fine-needle aspiration diagnosis have been reported.¹⁰⁻¹³ However, when the tumor presenting as soft or hard tissue mass in any localization, radical removal, en block or piecemeal resection, is the best therapeutic options. In this patient, complete tumorectomy, including synovectomy, of a Grade 3 (a and c) of the Sekhar's classification for temporal bone neoplasmas was the treatment of choice.¹⁴ To our knowledge, only one case has been reported of giant solitary synovial chondromatosis of the temporo-mandibular joint with intracranial extension.⁶

and surgically a well-recognized tumor and easy, total piecemeal resection was achieved in both cases. The follow-up period was 18 months without evidence of disease.

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