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Case Reports

Primary Synovial Chondromatosis of the Temporomandibular Joint

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Synovial chondromatosis is a benign tumour-like disorder characterized by the development of nodules of cartilage within the synovial membrane of articulating joints.¹ It is most often reported in the larger joints of the body, including the knee, hip, and elbow and ankle joints, although it would rarely appear to affect the temporomandibular joint (TMJ).² Synovial chondromatosis of the TMJ, first described by Auhausen in 1933,³ appears to be rare, the chief clinical features of this disorder being a pre-auricular swelling and pain.² Based on these clinical characteristics, many patients may seek help from otolaryngologists. Although a number of publications addressing the subject are available in the literature,^{4,5} most are restricted to specialized journals of dentistry and radiology, which may not be commonly read by otolaryngologists. We report on two cases of this disorder, which were first seen by otolaryngologists, including an attempted correlation of the imaging with surgical and histologic findings.

C-erb B-2, located on chromosome 17q21, is regarded to have a role in normal cell proliferation and development. It produces a 185 kD glycoprotein, which is a transmembrane receptor with tyrosine kinase activity and shares a 78% homology with the epidermal growth factor receptor.⁶ Overexpression of *C-erb B-2* has been reported in a wide variety of carcinomas but has only rarely been described in connective tissue tumours.^{7,8} To our knowledge, there is only one report of *C-erb B-2* staining in synovial chondromatosis.⁹ Furthermore, to our knowledge, there are no reported studies on *C-erb B-2* in TMJs with synovial chondromatosis. Therefore, we also immunohistochemically examine the expression of *C-erb B-2* in this report.

Case 1

A 39-year-old woman, referred by her otolaryngologist, presented with a 10-year history of a left-sided preauricular swelling that appeared to have originated from the parotid gland (Figure 1). Frank asymmetry of the patient's left face was evident, and, on physical examination, no trismus was noted, but a clicking sound over the left TMJ was noted when the patient closed her mouth. She also complained of frequent tinnitus emanating from her left ear. No history of trauma could be identified. A clinical impression of parotid gland neoplasm was considered at the time. Using mag-

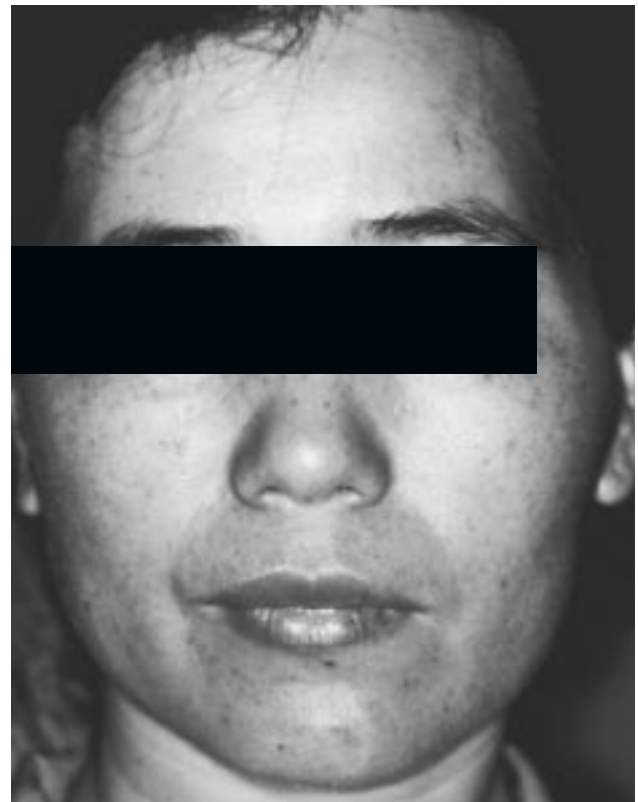


Figure 1 Case 1. A left-sided preauricular swelling with frank asymmetry in her left face can be noted.

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netic resonance imaging (MRI), a tumour-like lesion with an extracapsular extension over the left TMJ was suspected (Figure 2). Subsequently, under general anaesthesia, the left joint capsule was explored, and a tumour-like lesion was found extending from the synovial membrane into the superior joint space (Figure 3). The whole lesion, together with the attached membrane, was therefore totally excised. Following approximately 9 years of ongoing follow-up, normal mouth opening was noted, with no sign of recurrence of synovial chondromatosis.

Case 2

A 63-year-old woman was referred to our institution for pain in the left TMJ over the preceding 5-month period. Originally, she had visited an otolaryngologist, and medication was prescribed; however, the pain persisted. Clinical examination revealed tenderness to palpation in the left masseter muscle and temporalis muscle, with no apparent preauricular swelling. No obvious mouth-opening limitation was noted, the patient's maximal mouth opening being 34 mm; her anterior open bite was the cause of the complaint. When the patient opened her mouth, her mandible deviated noticeably to the right, with crepitus apparent (Figure 4). She otherwise remained fit and well, and there was no history of injury to the jaws.

Panoramic and transcranial radiographs revealed no obvious bony changes in either the glenoid fossa or the mandibular condyle. Widening of the left TMJ space was identified on computed tomography (CT) (Figure 5). There was apparent fluid accumulation



Figure 3 Case 1. An extracapsular mass can be noted over the left temporomandibular joint.

within the superior joint space as determined from a hyperintense signal on T₂-weighted images (Figure 6). These findings suggested a clinical diagnosis of TMJ dysfunction with joint effusion within the left TMJ.

To relieve the symptoms for the patient and to drain the joint effusion, a surgical approach to the left TMJ was adopted. Exploration of the left TMJ was performed under general anaesthesia. The left joint capsule was exposed and divided by a vertical incision. The disk and the superior joint space were then exposed. Immediately subsequent to opening the joint capsule, some transparent and viscous fluid issued forth from the upper joint space. Two small nodule-like masses over the superior joint space were incidentally noted and therefore removed. The patient recov-

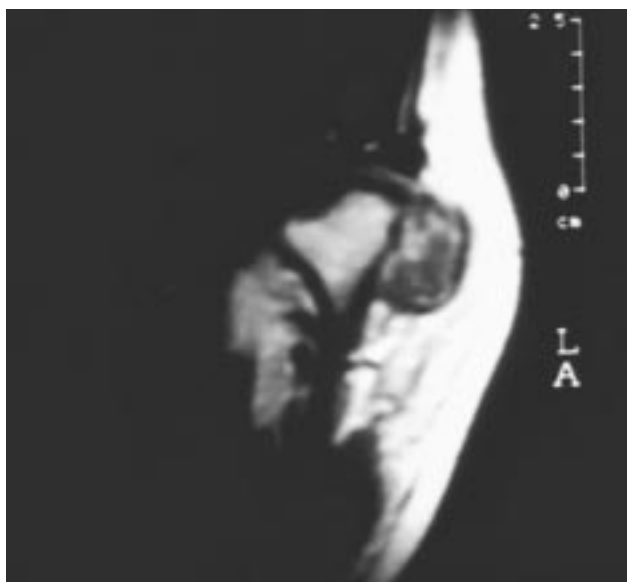


Figure 2 Case 1. Magnetic resonance imaging revealed a tumour-like lesion with extracapsular extension over the left temporomandibular joint.



Figure 4 Case 2. Anterior open bite with the mandible deviated to the right on mouth opening can be noted.



Figure 5 Case 2. Widening of the left joint space as identified by computed tomography.

ered well and was able to open her mouth to 45 mm 2 months postoperatively. The patient has been followed up regularly and has remained symptom free, with no recurrence, for about 2 years.

Histologic and Immunohistochemical Findings

The specimens were fixed immediately after surgery in 10% buffered formalin for approximately 24 hours and embedded in paraffin. Consecutive sections were

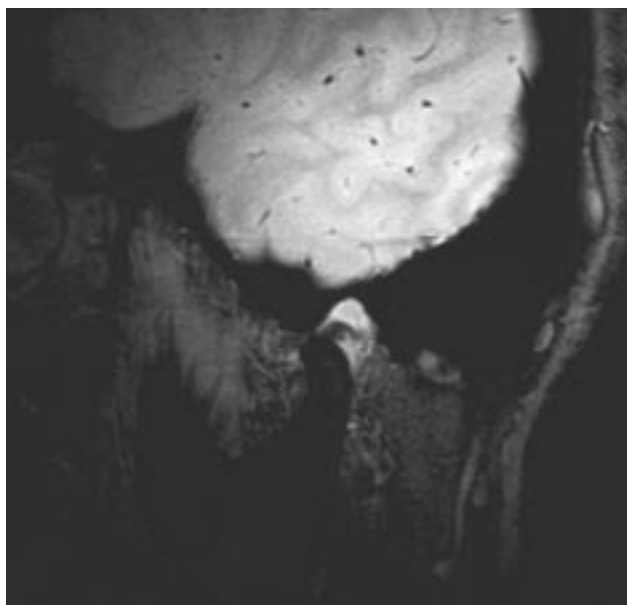


Figure 6 Case 2. Joint effusion within the left temporomandibular joint as identified by magnetic resonance imaging.

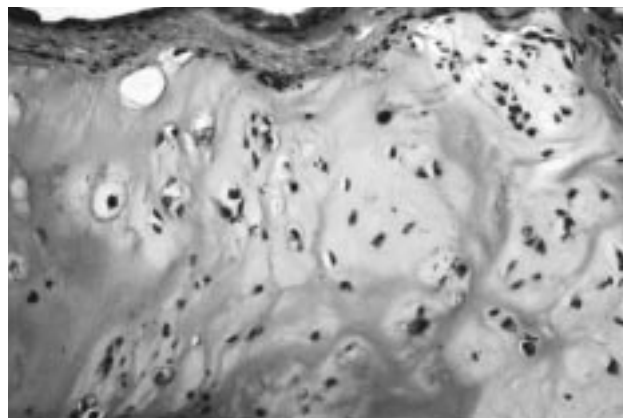


Figure 7 Case 1. Histologic examination of the excised tissue showing the cartilaginous tissue without cytologic atypia (hematoxylin-eosin stain; original magnification $\times 100$).

made and prepared for routine histologic study. The expression of C-erb B-2 was analyzed using a three-stage avidin-biotin immunoperoxidase technique. Immunostaining for C-erb B-2 was performed using rabbit polyclonal antibody (Dako, 1:100 dilution). The specificity of the antibody has been characterized.¹⁰ The sites of peroxidase binding were visualized as brown reaction products from diaminobenzidine reaction, followed by counterstaining with hematoxylin. Normal articular cartilage was included as control specimens. Positive controls included a known positive salivary gland tumour. Negative controls were included following the same procedure but with omission of the primary antibody and using an irrelevant rabbit immunoglobulin recommended by the manufacturer (Dako X903).

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Case 1

Histologic examination of the excised tissue was characteristic for synovial chondromatosis, consisting of chondrocyte-like cells covered with thin connective tissue, and the chondrocytes failed to display cytologic atypia (Figure 7). Immunoreactivity for C-erb B-2 was observed in less than 50% of the chondrocyte-like cells (Figure 8). Positive staining revealed positive, whereas negative control sections showed only background staining.

Case 2

Histologic examination of the surgically derived specimen revealed chondrometaplasia of the synovium of the TMJ (Figure 9); cellular atypia of the chondrocytes (Figure 10) was also noted. These histologic findings confirmed the diagnosis of a synovial chondromatosis of the left TMJ. Immunoreactivity for c-erb B-2 was observed in almost all of the chondrocyte-like cells (Figure 11). Positive staining revealed positive control sections, whereas negative control sections showed only background staining.

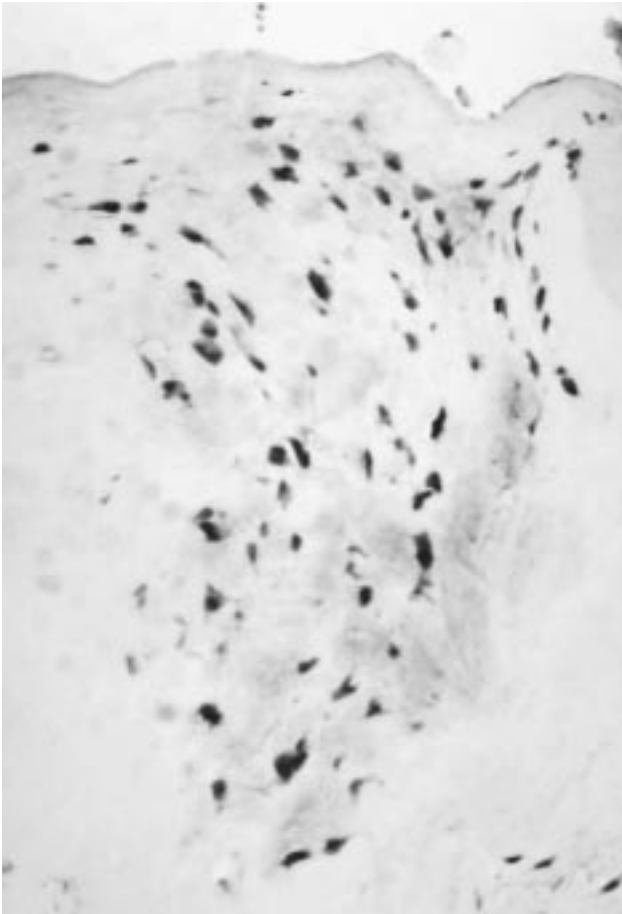


Figure 8 Case 1. Immunoreactivity for *c-erb B-2* was observed in less than 50% of the chondrocyte-like cells (avidin-biotin immunoperoxidase stain; original magnification $\times 100$).

Discussion

Synovial chondromatosis has often been considered to be a metaplastic rather than a neoplastic disease. It may be a primary condition, or it may occur secondarily to

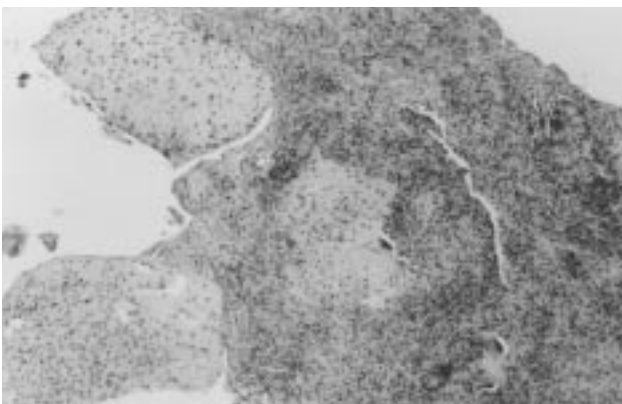


Figure 9 Case 2. Histologic examination of the specimen revealing chondrometaplasia of the synovium (hematoxylin-eosin stain; original magnification $\times 40$).

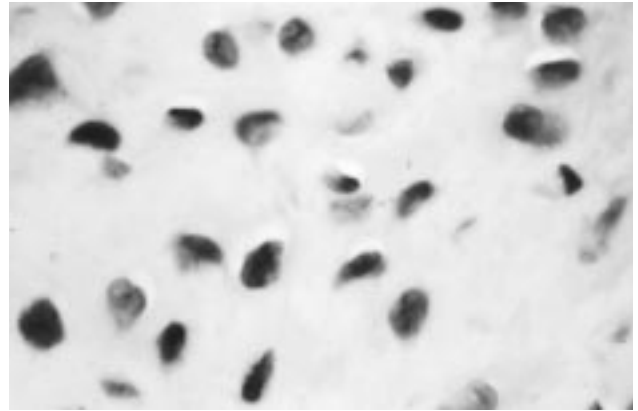


Figure 10 Case 2. Histologic examination of the specimen revealing cellular atypia among the chondrocytes (hematoxylin-eosin stain; original magnification $\times 100$).

other joint disorders, such as osteoarthritis, avascular necrosis, osteochondrosis dissecans, and/or rheumatoid arthritis.¹¹⁻¹³ No other joint diseases could be identified for our two cases; therefore, we believe that both cases are examples of primary synovial chondromatosis.

In this study, we observed positive staining in both cases of TMJ synovial chondromatosis but no positive staining in normal cartilage. Fewer than 50% of cells stained in one case and more than 90% in another case, but the staining was of strong intensity in both cases. This highlights the distinct difference between the cartilage found in primary synovial chondromatosis and normal cartilage. Positive staining has been demonstrated in primary synovial chondromatosis⁹ but has not been reported in its TMJ counterpart. Therefore, the present study may represent the first to demonstrate the expression of C-erb B-2 in TMJ synovial chondromatosis.

Today, the cause of this disease is still unknown, but trauma to the region has been implicated as a possible initiating factor.¹⁴ No trauma history, however, was identified for both cases presented in this report.

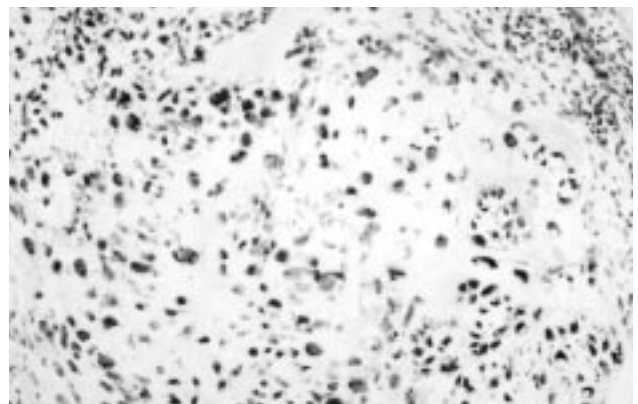


Figure 11 Case 2. Immunoreactivity for *c-erb B-2* was observed in almost all of the chondrocyte-like cells (avidin-biotin immunoperoxidase stain; original magnification $\times 40$).

On the other hand, transforming growth factor and tenascin have both been reported to be implicated in the genesis of synovial chondromatosis of the TMJ.¹⁵ Also, fibroblast growth factor 2 (FGF-2) and fibroblast growth factor receptor 1 (FGFR-1) have been found in chondrocytes in synovial chondromatosis of the TMJ, suggesting the possible roles of FGF-2 and FGFR-1 in this disorder of the TMJ.¹⁶ As mentioned before, *c-erb B-2* shares a significant homology with the epidermal growth factor receptor.⁶ Further studies are needed to elucidate the direct contribution of these growth factors, perhaps associated with *c-erb B-2*, to the pathogenesis of synovial chondromatosis of the TMJ.

Synovial chondromatosis of other joints has been reported to occur twice as often for males as is the case for females, with a mean age of onset of the condition occurring in the fifth decade.¹⁷ Within the TMJ, Fee and colleagues reported a significant female-to-male ratio of 4:1 and noted that the lesion was typically located on the right side of the body (right-to-left ratio of 4:1).¹⁸ In this report, both patients are female, substantiating the finding of Fee and colleagues¹⁸; however, in contrast, these two cases both revealed a lesion located in the left joint.

Our first case was referred to as a parotid neoplasm owing to the existence of a preauricular swelling, which, however, is also a common presenting symptom for synovial chondromatosis. If extracapsular extension of synovial chondromatosis exists, as was the case for our first patient, it can be mistaken for either a benign¹⁹ or a malignant²⁰ parotid neoplasm. Therefore, the correct differential diagnosis between a TMJ arthropathy and a parotid tumour is important. For our second case, both the presenting symptoms (muscle tenderness, mandibular deviation, and crepitus on mouth opening) and radiologic findings (joint effusion) mimic the presence of TMJ dysfunction. Therefore, if a patient presents with symptoms mimicking TMJ dysfunction, the possibility of synovial chondromatosis should also be considered. Also, it has been reported that because of its low incidence, the affected patients were not diagnosed during the initial visit and were delayed for appropriate diagnostic imaging and therapy at an early stage.⁴

Synovial chondromatosis has been classified into three phases: (1) early, with synovial chondrometaplasia but no loose bodies present; (2) transitional, with active synovial disease and loose bodies present; and (3) late, with loose bodies present, which may vary in size from < 1 to > 10 mm, but no apparent synovial disease.²¹ Based on the respective radiologic and histologic findings for our cases, we consider that our first case is representative of the first developmental stage, whereas our second case belongs to the transitional stage.

In 1977, Noyek and colleagues pointed out radiologic features of synovial chondromatosis in the TMJ, namely (1) a widening of the joint space, (2) a limita-

tion to joint movement, (3) irregularity of the joint surface, (4) the presence of calcified loose bodies, and (5) sclerosis or hyperostosis of the glenoid fossa and mandibular condyle.¹⁷ These radiologic features, however, are also commonly seen for cases of osteoarthritis involving the TMJ, apart from the presence of calcified loose bodies. Furthermore, other joint disorders, such as osteochondritis dissecans, intracapsular fracture, avascular necrosis, osteoarthritis, tuberculosis or pyogenic arthritis, rheumatoid arthritis, and neurotrophic arthritis, may also reveal the presence of loose bodies within the TMJ.^{22,23} Therefore, when these radiologic characteristics are not observed in their entirety, there may be some difficulty in obtaining an accurate imaging-based diagnosis of synovial chondromatosis. In our second case, with the absence of loose radiopaque bodies, a definitive diagnosis may not be easily obtained. However, with the presence of an abnormally large amount of synovial fluid on MRI, a diagnosis of synovial chondromatosis, as suggested by Kim and colleagues,⁵ must be considered.

Most authors believe that a synovial chondromatosis is a benign, tumour-like disorder, and subsequent malignant transformation is rare. Davis and colleagues reviewed 53 cases of synovial chondromatosis over a period of 30 years, reporting that nine patients suffered recurrence, including three cases that subsequently became malignant.²⁴ This suggests that primary synovial chondromatosis exhibits a potential for malignant change. Indeed, a case of synovial chondromatosis of the TMJ resembling chondrosarcoma has recently been reported.²⁵ Multiple and often short-interval recurrences of synovial chondromatosis should alert the physician to the possibility of synovial chondrosarcoma.²⁶ For our first case, there was no sign of recurrence after a long-term (9 years) follow-up period. For our second case, owing to the presence of cellular atypia among the chondrocytes, the possibility of chondrosarcoma based on the histopathologic features should be considered; however, because neither invasive nor metastatic findings were noted, we made a diagnosis of synovial chondromatosis. Although there has been no sign of recurrence of this condition for our second patient to date, we do plan to carefully follow this patient for the long term.

The removal of the loose bodies and of the involved membrane from the diseased TMJ is believed to be sufficient to control the disease for the majority of cases, although the treatment of choice remains somewhat controversial, with many authors advising synovectomy and removal of loose bodies either by arthroscopy or by open operation. We adopted the open surgical procedure to remove the lesion for the first patient because the lesion had already exhibited an extracapsular extension, whereas to relieve the symptoms for our second patient and to drain the joint effusion, the patient was subjected to open surgery.

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