Synovial Chondrosarcoma Arising in Synovial Chondromatosis of the Temporomandibular Joint

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Abstract Synovial chondromatosis of the temporomandibular joint is rare. Even less commonly documented is the progression of synovial chondromatosis to a synovial chondrosarcoma. The aim of this paper is to present only the third case of synovial chondrosarcoma of the temporomandibular joint. Distinction between these two entities by histology alone is extremely difficult and even though it is advised that the definitive diagnosis should be based on clinical, radiographic and histological evidence, this has proved not to be so simple. The patient, a 63 year old female presented with a swelling associated with her left temporomandibular joint. CT and MRI scans confirmed the presence of a periauricular chondroid mass. Fine needle aspiration biopsy revealed an atypical chondroid lesion that was suspicious for a chondrosarcoma. The left temporomandibular joint and surrounding tissues were resected after further imaging and extensive clinical, radiological and cytologic consultations. A diagnosis of synovial

chondrosarcoma arising in synovial chondromatosis was made.

Keywords Chondrosarcoma · Mandibular condyle · Synovial chondromatosis · TMJ

Introduction

Synovial chondromatosis or chondrometaplasia was always considered to be a benign, non-neoplastic monoarticular process that very uncommonly involves the temporomandibular joint [1–6]. It has however recently been suggested that it may be a benign neoplasm which presents as multiple hyaline cartilage nodules typically within subsynovial tissues [7]. Two forms of the disease have been recognized: primary and secondary. In the primary form, which is uncommon, the aetiology is not known, however a response to repetitive, low-grade trauma has been proposed. Secondary synovial chondromatosis is more common and arises as a result of an inflammatory or non-inflammatory arthropathy [2]. As a result, the hip and knee are the preferred sites of involvement. Axhausen [8], in 1933, described the first report of synovial chondromatosis affecting the temporomandibular joint (TMJ). This condition is characterised by foci of metaplastic cartilage or osteocartilagenous nodules within the synovial membrane or lying free within the joint space. Reviews of the literature until 2000 revealed at least 79 cases of synovial chondromatosis that have involved the TMJ [1, 6]. Since then several additional cases of synovial chondromatosis of the TMJ have been reported [3–5].

Synovial chondrosarcoma is a very rare malignancy that arises in synovial chondromatosis or de novo within the synovial membrane. Only 34 cases had been reported in the literature by 2003 [9, 10]. The incidence of this rare

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sarcoma is unknown and most reports have been of single cases [11–14]. The knee and hip joints have been most frequently affected and to the best of our knowledge only two previously reported cases have been described involving the TMJ [15, 16].

Case Report

A 63 year-old female was referred with swelling associated with her left TMJ. The lesion had been slowly enlarging but was otherwise asymptomatic. There was no history of facial trauma or prior radiation. The remainder of her medical history was essentially unremarkable. Examination revealed minimal facial asymmetry with a diffuse swelling overlying the left TMJ. Mouth opening and facial nerve function was normal. There was no regional lymphadenopathy.

CT scans, MRI and plain radiographs showed a mass which demonstrated features consistent with a chondroid lesion (Fig. 1a, b). A soft tissue component was observed both anterior and posterior to the neck of the left mandibular

condyle with erosion of the posterior aspect of the condylar neck being identified. There were no peri-articular calcifications. The chondroid lesion appeared to be centered within the peri-articular soft tissues with sclerosis of the posterior aspect of the neck of the condyle being observed rather than a permeative pattern of bone involvement. The attending radiologists had differing opinions as to the nature of the lesion and the differential diagnoses considered included a chondrosarcoma and synovial chondromatosis.

After imaging, the patient underwent an ultrasound guided FNA biopsy of the lesion which yielded abundant material that showed an atypical chondroid lesion that was highly suspicious for a chondrosarcoma (Fig. 2a, b). Synovial chondromatosis however could not be excluded.

After extensive clinical, radiographic, cytologic review and discussion, it was decided to resect the lesion together with the involved condyle. At operation the left TMJ and portion of the zygomatic arch were resected together with a surrounding soft tissue margin. The upper division of the facial nerve was divided to allow access and then repaired after the resection. The surgical defect was repaired with a

Fig. 1 **a** MRI scan—coronal slice demonstrating lobulated chondroid lesion surrounding the condylar neck both medially and laterally. **b** MRI scan—axial slice demonstrating lobulated chondroid lesion surrounding the condylar neck both anteriorly and posteriorly

Fig. 2 **a** FNAB specimen demonstrating classical lobulated appearance of 'clones' of chondrocytes. **b** FNAB specimen with focal atypia of chondrocytes

soft tissue free flap and a rib graft to reconstitute the zygomatic arch. No attempt was made to reconstruct the joint. The patient was kept in intermaxillary fixation for some weeks post-operatively.

The specimen measured $50 \times 55 \times 40$ mm and consisted of a large soft issue mass which surrounded the left TMJ. It included the parotid gland, ascending ramus, coronoid process, condylar head, meniscus as well as the glenoid fossa and the zygomatic bone (Fig. 3).

Histologic examination showed parotid gland parenchyma with adjacent fibro-adipose tissue covered by synovium, bone, cartilage and the mandibular condyle. Variably sized nodules of hyaline cartilage exhibiting clustering of chondrocytes were observed within the sub-synovial tissue, characteristic of synovial chondromatosis

Fig. 3 Sagittally transected specimen following decalcification. The lobulated chondroid tumour surrounds the neck of the condyle with pressure erosion of the posterior cortical bone

(Fig. 4a, b). Some nodules extended into the surrounding soft tissue encircling and encompassing a large nerve (Fig. 4b). In several areas, peripheral cellularity was present in which the peripheral chondroid cells showed a marked degree of nuclear variability in shape and size with multinucleated cells including binucleate and trinucleate forms being observed (Fig. 5). Many of the component cells had large, irregular vesicular nuclei with prominent nucleoli and occasional mitotic figures were noted (Fig. 6). Myxoid change of the matrix was present focally (Fig. 7). Definite spindling of the tumour cells was not observed.

Examination of the decalcified mandible showed erosion and remodeling of the posterior neck of the condyle with a pushing margin as a result of the synovial chondromatosis. There was no convincing evidence of bone penetration and permeation by the atypical chondroid tissue (Fig. 4a). A diagnosis of synovial chondrosarcoma arising in synovial chondromatosis was made. The patient then received adjuvant radiotherapy and at present is well and free of disease.

Discussion

The histological diagnosis of low grade chondroid lesions of the bones is fraught with difficulty. The diagnosis of malignancy in soft tissue chondroid lesions is even more so as considerable cellularity including binucleation is common in benign lesions involving the soft tissue such as synovial chondromatosis and soft tissue chondroma [17]. As such the diagnosis of malignancy on FNA of chondroid lesions is not recommended. In all cases, correlation with relevant radiographs and scans is necessary in order to establish a definitive diagnosis [18].

Both benign and malignant chondroid lesions of the jaws and facial bones are extremely uncommon [18] and lesions involving the TMJ are even less frequently encountered. It has even been suggested by Saito et al. [18]

Fig. 4 **a** Lobulated chondroid tumour surrounding the neck of the condyle with pressure erosion of the posterior cortical bone. **b** Typical lobules of synovial chondromatosis with 'clones' of chondrocytes interspersed between nerve fibres

Fig. 5 Typical synovial chondromatosis above with area of sarcomatous transformation

Fig. 6 Sarcomatous transformation with pleomorphic chondrocytes with nuclear hyperchromatism

Fig. 7 Myxoid degeneration within the malignant component of the tumour

that any clinically significant cartilaginous mass in the region of the jaw and face should be considered to be malignant. Lesions containing chondroid that involve the TMJ include post traumatic arthritis, chondromyxoid fibroma, osteochondroma, synovial chondromatosis, chondrosarcoma and chondroblastic osteosarcoma.

Arthritic changes are the most common pathologic conditions involving the TMJ [19]. Degenerative osteoarthritis and rheumatoid arthritis would be the most common causes however they would not present a problem in the histological differential diagnosis of an atypical chondroid lesion because of their characteristic radiological findings, gross changes and histological features. Post-traumatic arthritis of the TMJ shows replacement of much of the articular surface and the condylar osseous cortex by hyaline cartilage with irregular distribution and arrangement of the chondrocytes. There is also intermingling of the cartilage with the underlying trabecular bone of the condylar head [20].

Chondromyxoid fibroma is a rare benign neoplasm considered to be of cartilaginous origin. Approximately 20 cases involving the jaws have been documented with 16 cases in the mandible and 4 cases that involved the maxilla. Histological diagnosis may be challenging on curetted material where the lobular architecture is lost and cellular atypia which may result in overdiagnosis as a chondrosarcoma. Review of the radiographs may help exclude a chondrosarcoma in which the classical sclerotic margins would not be expected [2]. These lesions are usually centered in bone and not in the soft tissue.

Chondromas are extremely rare in the jaws and facial bones. The diagnosis of enchondroma should only be made when the lesion is a small incidental finding [18] since larger lesions are more likely to be malignant. They are centered in bone. The histological features of an enchondroma and a low-grade chondrosarcoma are similar and differentiation of one from the other can be difficult in the absence of demonstrable permeation [17]. In most cases radiographic evidence of aggressive features including cortical destruction allows for a diagnosis of low-grade chondrosarcoma to be made.

Osteochondromas of the TMJ have been reported [2, 6]. They are rare benign usually exophytic outgrowths of cortical and medullary bone with an overlying cartilaginous cap. These lesions probably represent misplaced epiphyseal plates on the surface of the bone rather than being a true neoplasm [21]. In a review of the literature by Barnes et al. [2], 39 cases involved the mandible with the preferential sites being the coronoid process (54 %) and condyle (41 %). This is a reasonable expectation since these are the sites in the mandible that are of embryological cartilage derivation. On imaging, continuity of the base and medullary cavity of the affected bone is usually demonstrable. This was not noted in our case. The histological features are characteristic and generally will not be confused with other chondroid lesions. The bone is covered by the cap of hyaline cartilage in which the chondrocytes have a linear orientation similar to a normal appearing epiphysis [22]. Occasional binucleate chondrocytes may be observed, however this should not be misinterpreted as a feature of

malignancy. Separate soft tissue nodules with a thick cartilage cap is a feature of periosteal chondrosarcoma in which pleomorphic cells as seen in our case may be identified. This was considered.

Chondrosarcomas of the jaw and facial bones are malignant neoplasms with hyaline cartilage differentiation and are extremely uncommon [23]. In a review from the Mayo Clinic they accounted for only 3 % of all chondrosarcomas. Most of these lesions presented in the nasomaxillary complex with only 11 % involving the mandible [18]. These tumours are characterized by a lobulated growth pattern of hyaline cartilage that showed bone permeation. The cartilage showed myxoid degeneration, hypercellularity, nuclear pleomorphism and binucleation of the chondrocytes. They are usually centered in bone and will show aggressive imaging.

The most frequently encountered pathology in the jaws showing chondroid differentiation is the chondroblastic variant of osteosarcoma. Chondroid differentiation in osteosarcomas is more commonly observed in lesions involving the jaw when compared to other sites and as a result many chondrosarcomas of the jaws have probably erroneously been misdiagnosed [18]. This differentiation of osteosarcoma from chondrosarcoma has important clinical implications, since although the most effective treatment for both types of sarcoma is radical surgical resection, osteosarcomas are also treated with chemotherapy as part of multimodality therapy. In our case features of osteosarcoma were absent both on imaging and on histology.

Separation of synovial chondromatosis from chondrosarcoma is a real diagnostic challenge both cytologically and histologically and in this case even more difficult as the malignant transformation existed only focally within the lesion [22]. The two entities overlap in their imaging, cytologic and architectural features and since the areas of progression were only observed focally, they may have easily been missed. Correct interpretation of synovial chondromatosis is a common pitfall in orthopaedic pathology particularly if interpretation is performed on cytologic and histological grounds only. Malignant transformation of synovial chondromatosis to a chondrosarcoma is rare [9] but the reported risk of progression to malignancy may as high as 5 % [24]. The diagnosis is very challenging and it is necessary to confirm that the location or centeredness of the lesion is within the soft tissues and to exclude an underlying osteochondroma with malignant transformation.

Bertoni et al. [14] have proposed some useful histological features that may assist in differentiating synovial chondromatosis and synovial chondrosarcoma from one another. These include atypical chondrocytes arranged in sheets, myxoid change of the matrix, mitotic figures, crowding and spindling of nuclei at the periphery of the

nodules, necrosis and, in particular, permeation of bony trabeculae. In our case, most of these criteria were met although definite permeation of bone was not evident. Sperling et al. [9] have also attempted to resolve this diagnostic dilemma with the aid of immunohistochemical staining techniques which included Bcl2, Ki-67, p27, p16 and p53. Their preliminary findings have shown that staining for Bcl2 may help differentiate the lesions from one another where chondrosarcomas show a loss of expression of Bcl2.

Histological distinction of synovial chondromatosis and its progression to a synovial chondrosarcoma is problematic and even with extensive multidisciplinary input, definitive diagnosis may still prove elusive due to the rarity of these conditions.

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