Juxta-Articular Myxoma: An Unusual Benign Mesenchymal Lesion, Readily Mistaken for Malignancy

Abstract:

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Abstract

Myxomas are benign tumours of mesenchymal origin. We describe the 1st reported case of paraspinal juxta-articular myxoma. Juxta-articular myxomas show increased cellularity and distinction from cellular myxoma is required. The differential also includes malignant myxofibrosarcoma. For patient prognosis and management it is essential to separate these entities. Complete surgical excision is the mainstay of treatment as local recurrences may occur.

Introduction

We describe a paraspinal juxta-articular myxoma. Myxomas are uncommon benign tumours of mesenchymal origin, mostly intramuscular in location. Juxta-articular myxoma, a variant of myxoma occurring around large joints, especially the knee (88%), is consideredbenign[1,2]. Reported sites include shoulder, elbow, ankle and hip joints[2]. To date, no case involving paraspinal joints has been reported. The main histological differentials are cellular myxoma and low grade myxofibrosacroma.

Case Report

A 55 year old female presented with a palpable, otherwise asymptomatic neck mass. It was present for 4 years, enlarging slowly over that time. There was no history of trauma. MRI of the cervical spine revealed an ovoid, loculated, partially cystic lesion at the level of C5 to C8, intimately associated with the transverse processes of the vertebral bodies and lying between the vertebral bodies and the sternocleidomastoid muscle. Intraoperatively, a 4 cm non-encapsulated mass was identified deep to cervical fascia adjacent to cervical vertebrae posterior to the upper end of the left sternocleidomastoid muscle. This did not appear radiologically or intra-operatively to arise from muscle. This mass was excised with clear margins. Multiple pieces of soft tissue were received in the laboratory. Microscopic sections revealed a poorly circumscribed cellular lesion in-between skeletal muscle (Figure 1). The tumour was composed of ovoid and spindle cells within a myxoid stroma. Myxoma describes any bland hypocellular gelatinous neoplasm[3]. When intramuscular, one expects a paucicellular lesion with round/stellate cells intermixed with myxoid extracellular stroma containing sparse capillary sized blood vessels[4]. Nuclear pleomorphism or mitoses are absent. Juxta-articular myxoma, is more cellular with increased vascularity[4]. Distinction from a cellular myxoma is based on location adjacent to large joints and the lack of a GNAS1 mutation[5]. Distinguishing between juxta-articular myxoma and low grade myxofibrosarcoma requires assessment of nuclear pleomorphism, hyperchromasia, and the presence of mitoses or necrosis[3].

Given the absence of mitoses, necrosis, hyperchromasia or nuclear pleomorphism in this case (Figure 2), low grade myxofibrosarcoma was excluded. This myxoma was cellular with admixed vessels and, given the radiologic and intraoperative confirmation of a lesion arising from the cervical vertebrae,a diagnosis of juxta-articular myxoma was made. The degree of cellularity, whilst unusual in a myxoma, is well described in juxta-articular myxoma[5]. This case was referred to Professor Christopher Fletcher, a world expert on soft tissue pathology, who confirmed the diagnosis.

Discussion

Myxomas most commonly occur between the ages of 40 and 60 years, with a female predilection[6]. They originate from primitive mesenchymal stem cells with differentiation towards altered fibroblasts that have lost the ability to produce collagen. Instead they produce hyaluronic acid and immature collagen[7]. They occur in many locations including heart, bones, genitourinary tract, skin, retroperitoneum, intestine, joints and most commonly skeletal muscles[3]. Myxomas are non-encapsulated, and may infiltrate surrounding tissues, however they do not metastasize[8]. Myxomas are slow growing tumours presenting as a painless mass or compression of surrounding structures[9]. Plain films are often normal or show a non-specific soft tissue mass. Ultrasound reveals a hypoechoic mass containing fluid filled clefts or cysts. CT shows a homogeneous low density mass. MRI shows a low intensity mass on T1 weighted images and high signal intensity on T2[7]. The imaging characteristics in both intramuscular myxoma and juxta-articular myxoma will be similar, except for the relationship to adjacent structures such as muscle and joints[6]. In cases of myxofibrosacroma, a more infiltrative growth pattern is seen on CT[8].

Most tumours are ovoid/globular with a glistening grey white appearance[9]. Microscopically, myxoma is hypocellular composed of undifferentiated stellate cells with an irregular meshwork of reticulum fibres in a matrix of hyaluronicacid-containing mucoid[10]. Cellular myxoma is more cellular with increased vascularity[4]. Juxta-articular myxomas are similar tocellular myxoma but lack a GNAS1 mutation[5], and are associated with large joints. Distinction from low grade myxofibrosacroma requires exclusion of nuclear pleomorphism, hyperchromasia, mitoses and necrosis[3]. These changes can be focal, and lead to difficulty, on small biopsy specimens. Full surgical excision is the treatment with core biopsy inappropriate. Myxomas do not metastasize however local recurrence is described in cases with incompleteresection[8]. Juxta-articular myxomas are more prone to recur[1].

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