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CASE REPORT

Granular Cell Tumour of the Deltoid Muscle

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ABSTRACT

Granular cell tumour is an uncommon neoplasm that generally has a benign behaviour. It typically occurs in the head and neck. The authors describe an unusual location of this tumour within the deltoid muscle of the arm.

Key Words: *Deltoid muscle, Granular cell tumour, Magnetic resonance imaging, Myoblastoma, Schwann cell*

INTRODUCTION

Granular cell tumour is a rare neoplasm that is so-named due to its arrangement of nests of polyhedral cells with abundant granular eosinophilic cytoplasm. These tumours typically present as a solitary mass. The vast majority are benign, with only approximately 2% of lesions demonstrating malignant histology and behaviour.[1] Granular cell tumours most commonly occur in the head and neck, with a particular predisposition for the tongue. Other common sites include the respiratory tract, the breast, and the gastrointestinal tract; however, virtually any body site may be affected. When granular cell tumours occur in other parts of the body, they typically involve the subcutaneous tissues. The authors present a case of benign granular cell tumour in a distinctly unusual location within the deltoid muscle of the arm. The magnetic resonance imaging (MRI) features of the lesion are discussed, along with a review of the current literature.

CLINICAL DETAILS

A 41-year-old female patient was referred to hospital by her family physician for further evaluation of a mass in the anterior part of her right shoulder. She had initially noticed a lump in this area 18 months prior to her present referral. At that time, the lump was pea-sized and not felt to be of clinical consequence. However, in the interim the mass had grown significantly in size. There was also mild discomfort present during certain arm movements such as arm abduction. There was no pain at rest and there were no systemic symptoms. She had no significant past medical or surgical history and was not on any medications. There was no family history of tumours.

On physical examination, there was evidence of a mass measuring approximately 3×3 cm in size located anterior to the shoulder joint. The mass appeared to be deep in position and related to the deltoid muscle. It did not appear fixed and was mobile when the arm was in a relaxed position. There was no palpable regional adenopathy and the rest of the physical examination was unremarkable. Routine laboratory investigations were all normal. A plain radiograph of the upper arm and right shoulder were normal with no osseous abnormality or soft tissue calcification seen. MRI was therefore arranged for further evaluation of the mass.

Axial and oblique coronal imaging pre- and post-Gadolinium was performed over the right shoulder region. This demonstrated a mass within the anterior portion of the deltoid muscle corresponding to the clinically palpable lesion. The mass measured $4 \times 2.8 \times 2.3$ cm in size. The mass involved most of the thickness of the deltoid muscle and abutted the humeral shaft posteriorly. There was no evidence of osseous invasion or signal change within the bone marrow. The mass was remote from the neurovascular bundle. Parts of its margins were well defined, but this was not uniform, with portions of

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the margins appearing less well defined. The mass was almost isointense to muscle on unenhanced T1-weighted images (Figure 1), with mild enhancement following the use of intravenous Gadolinium (Figure 2). The mass was of high signal on T2-weighted imaging (Figure 3) and short tau inversion recovery sequences (Figure 4). No discernable calcification, fat or blood products were detected within the mass. No other lesions were seen. The findings were not specific to a certain diagnosis and a biopsy was therefore arranged. A computed tomography (CT) scan of the chest prior to the biopsy was normal with no evidence of metastatic disease.

Two 18 gauge core biopsies were obtained from the mass under direct ultrasound guidance, and subsequently the tumour was surgically removed. The surgically resected specimen demonstrated a poorly defined 4.0 × 2.8 × 2.3 cm tan firm tumour mass within the deltoid muscle. The mass was noted to extend to the resection margins of the specimen at multiple points. Microscopically the biopsy material and sections of the resection specimen were identical. The tumour was composed of ill-defined cords and nests of oval epithelioid cells that had abundant granular eosinophilic cytoplasm (Figure 5). The tumour cell nuclei were central and vesicular.

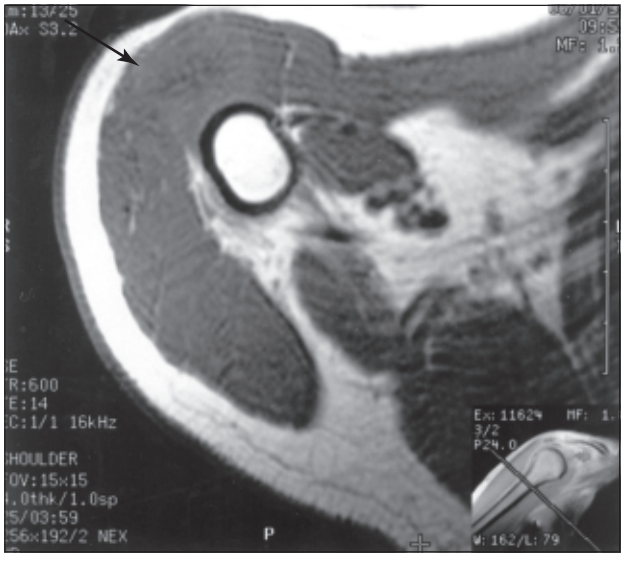

Figure 1. Axial non-contrast T1-weighted image through the right upper arm (TR 600, TE 14) demonstrates a mass within the anterior portion of the deltoid measuring 2.8 × 2.3 cm in size (arrow).

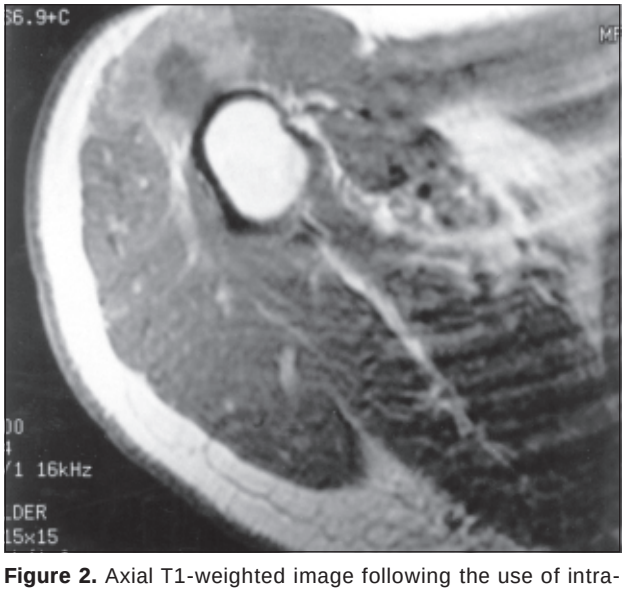

Figure 2. Axial T1-weighted image following the use of intravenous Gadolinium (TR 600, TE 14) shows enhancement of most of the periphery of the mass with only the central part of the mass remaining isointense to muscle.

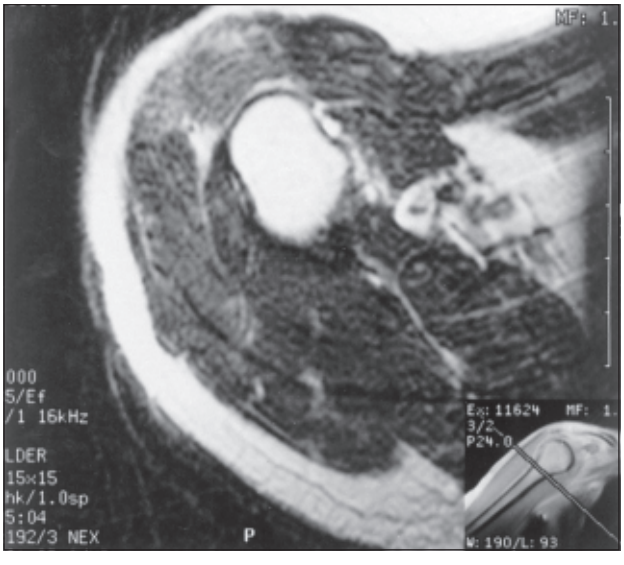

Figure 3. Axial FSE T2-weighted image (TR 4000, TE 85/Ef) demonstrates high signal within the mass.

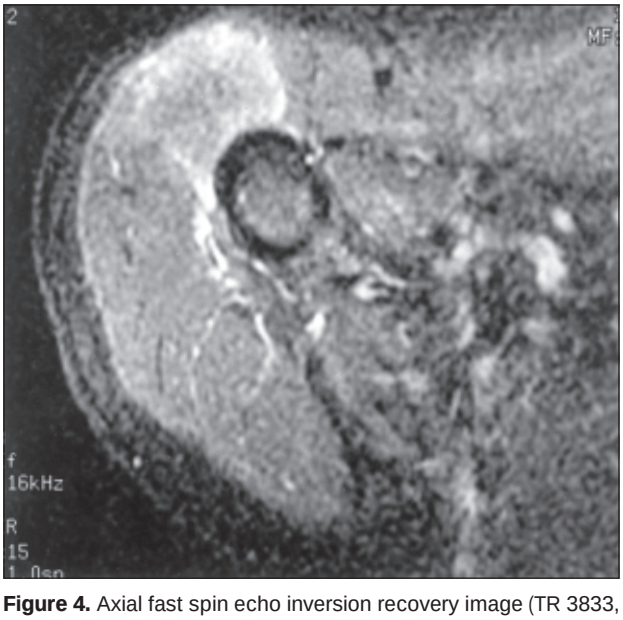

Figure 4. Axial fast spin echo inversion recovery image (TR 3833, TE 34/Ef, TI 150) demonstrates high signal within the mass with no loss of signal to suggest fat within the lesion. No abnormal marrow signal is seen within the humerus.

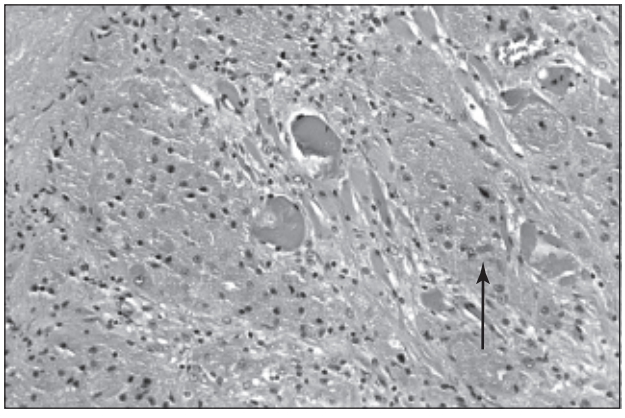

Figure 5. Characteristic appearance of nests of large polyhedral cells with abundant granular cytoplasm (arrow). The tumour cells surround and infiltrate between mature skeletal muscle cells. (Haematoxylin and eosin stain, × 55 original magnification)

Occasional cells demonstrated small nucleoli. Mitoses were scant and there was no evidence of necrosis. Spindle-shaped tumour cells, fascicular growth, or a high mitotic rate were not identified. The resection margins of the specimen were microscopically positive for tumour.

In view of the histologically positive resection margins and the inherent risk of tumour recurrence, the patient was subsequently followed up closely. At 1 year follow-up, the patient remains well with no evidence of recurrence.

DISCUSSION

Granular cell tumour is a very unusual tumour that was first described by Weber in 1854. It was classified by Abrikossoff as a pathological entity in 1926.[2] The origin and nature of granular cell tumour have been debated since its original description. Although the original theories proposed that granular cell tumours were derived from immature skeletal muscle cells, subsequent investigations using electron microscopy and immunohistochemistry have proven that these tumours exhibit Schwannian differentiation.[3] Most granular cell tumours are poorly circumscribed nodules, typically measuring less than 3 cm in maximum diameter. However, there is a range of sizes and appearances, and sizes of over 10 cm have been described in the literature.[4] Growth tends to be very slow except for the rarer malignant granular cell tumours, in which growth is generally much more rapid.

Of the reported cases, approximately 30% arise in the oral cavity, with the tongue being the predominant site of occurrence.[5] Another 30% originate in the skin and subcutaneous tissues, while 15% are localised in the breast, and 10% in the respiratory tract.[1] The tumour has also been described in skeletal muscles, gallbladder, urinary bladder, female genitals, and the peripheral and central nervous system.[4] The average age of patients is 32 years, with a typical age range of 15 to 60 years. Males are affected twice as commonly as females.

Conventional granular cell tumours are benign neoplasms. Malignancy occurs in less than 2% of patients. Grossly, granular cell tumours appear as poorly demarcated homogenous, greyish-white or tan nodules. Microscopically, the benign tumours are characterised by nests and cords of large polyhedral cells with centrally located, small, evenly-stained nuclei. As their name suggests, there is abundant granular eosinophilic cytoplasm.[6] The tumours are typically poorly circumscribed, with the granular cells trailing off into surrounding tissues from the main mass of cells. Typically, when located adjacent to an epithelial lined surface, granular cell tumours elicit marked overlying epithelial hyperplasia. Sometimes this epithelial proliferation is so marked that it may resemble an infiltrating carcinoma. Malignancy is diagnosed by a combination of histological findings, including cellular pleomorphism and elevated mitotic activity, and clinical malignant behaviour. Clinical suspicion of malignancy should be higher if the lesion is large or growing rapidly, or if there is evidence of distant spread.[7] Not only do malignant lesions tend to be larger and more aggressive in behaviour, but they also tend to occur at different sites than the benign variety. While oral lesions are the primary site for benign tumours, malignant lesions are most common within the chest wall, followed by the thigh.[8]

Most granular cell tumours present as a solitary painless lesion. Rarely, ulceration may occur, particularly in the buccal surface of the mouth. These tumours are often discovered incidentally. Nevertheless, pain, tenderness or sensitivity may be referred from adjacent structures that are influenced by the mass. Not uncommonly, minor trauma to the body part containing the lesion, or trauma to the lesion itself, is what first brings the lesion to the patient's attention. Approximately 10% of patients demonstrate multifocal disease, frequently involving subcutaneous, submucosal, and visceral tissues, either synchronously or metachronously.[9] Multifocal disease is more common in black patients.[4] There is a higher incidence of malignancy when multiple body sites are involved.[10]

Radiologically, granular cell tumours are best evaluated with MRI. Typically the tumours are slightly hypointense on T1-weighted sequences, and show homogenous contrast enhancement after intravenous injection of Gadolinium.[11] On T2-weighted sequences, tumours generally show a heterogeneous increased signal. While the MRI tissue characteristics are non-specific, MRI does allow precise localisation of the tumours and is invaluable in the preoperative evaluation of patients with a granular cell tumour. On CT, tumours appear solid, and show homogenous enhancement. Ultrasound may sometimes be useful for evaluating these lesions.

The treatment advocated for granular cell tumours is excision with wide margins.[12] The margins should be sufficiently wide to obviate possible local recurrence. Recurrence after resection is rare for benign lesions.[6] Malignant lesions, however, commonly recur, both at a local level and with local lymph node metastatic disease.[8] Because of the propensity of malignant lesions to local recurrence and regional lymph node involvement, some authors have recommended regional lymph node dissection when tumours display rapid growth or exceed 4 cm in diameter.[13] Where there are clinical or histological features of malignancy, patients should undergo a more comprehensive imaging evaluation, including CT of the chest, in order to evaluate for metastatic disease. Radiotherapy and chemotherapy have been used in the past when malignant disease was present. Variable success has been reported and the effectiveness of these treatments is anecdotal and unproven.

In summary, granular cell tumour presenting in the deltoid muscle is distinctly unusual. MRI was useful in this case, both to confirm the presence of a mass, and to detail its location and relationship to other structures prior to surgery. Core biopsy was, however, necessary to make the diagnosis, as the imaging findings of granular cell tumour are often non-specific. While radiologists evaluating such a mass are not likely to make the diagnosis on imaging findings alone, it is important for them to be aware of the diagnosis and the generally good prognosis that it carries.

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