Case Report

Granular Cell Tumour of the Deltoid Muscle: A Rare Entity in The Musculoskeletal System

Chetan S Chaudhari*, Shailesh S Vartak, Urmi S Chakravarty, Manisha S Khare, Prashant V Kumavat

Dept. of Pathology, Lokmanya Tilak Municipal Medical College & General Hospital, Sion, Mumbai, India

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ABSTRACT

Granular Cell Tumours are a rare mesenchymal soft tissue tumours that arise throughout the body and are believed to be of neural origin. They are so named because composition of characteristic nesting pattern of polyhedral cells with abundant granular eosinophilic cytoplasm. They often present as asymptomatic, slow-growing, benign, solitary lesions but may be multifocal. 1-2% of cases are malignant and can metastasise. Granular cell tumour commonly occurs in head and neck,with a particular predisposition for the tongue. The authors describe an unusual location of this tumour within the deltoid muscle of the arm. A 55 year old female presented with mass and tenderness at the distal end of deltoid site since 2 years. MRI showed a mass within deltoid muscle, 5x3x2 cm. Intraoperative frozen and later surgically resceted specimen showed histopathological features of benign granular cell tumour, at unusal site and a rare entity in the musculoskeletal system , witin deltoid muscle.

***Corresponding author:**
Dr. Chetan S Chaudhari, 11, Jagannath Darshan, M.D. Keni Road near RBI Quarters, Bhandup Village (E), Mumbai, Maharashtra, India
Phone: +91 - 9819133606
E-mail: drchetanchaudhari26@yahoo.co.in

Introduction

Granular cell tumour (GCT) is an uncommon benign neoplasm, first described by Abrikossoff in 1926 [1] The tumour was initially called 'granular cell myoblastoma' due to its possible proposed origin from skeletal muscle. Granular cell tumour is so-named due to its arrangement of nests of polyhedral cells with abundant granular eosinophilic cytoplasm. Various theories on the origin of GCT have subsequently been proposed, including its origin from striated muscle and histiocytes and a neural origin. Granular cell tumours can affect any organ or region of the body. Most GCTs occur in the head and neck region, especially in the tongue, cheek mucosa, and palate. [2] Other common sites include the respiratory tract, the breast, and the gastrointestinal tract. These tumours typically present as a solitary mass. The vast majority are benign, with only approximately 2% of lesions demonstrating malignant histology and behaviour. [3] The authors present a case of benign granular cell tumour in a distinctly unusual location within the deltoid muscle of the arm with only a sparse of such cases reviewed in literature. Clinical features, magnetic resonance imaging (MRI) features with characteristic histopathological features of the lesion are discussed, along with a review of the current literature.

Case Report

A 55 year old female presented with mass and tenderness at the distal end of deltoid site since 2 years. 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 Clinical examination, there was evidence of a mass measuring approximately 5x3 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.All other routine hematological and biochemical investigations were within normal limits.Plain radiograph of shoulder showed no osseous abnormality.

MRI examination showed a solitary poorly circumscribed mass, measuring 5x3x2 cm within deltoid muscle at its distal end. The mass was hypointense to muscle on unenhanced T1-weighted images and away from neurovascular structures.No intratumoural calcification, hemorrhages or necrosis was seen.(Fig1) Preoperative FNAC procedure for mass was not performed. Intraoperative frozen section was sent.Patient underwnt surgical excision of mass, which was poorly circumscribed, lobulated,homogenous, and was greyish tan (Fig2). It showed on frozen and on histopathology poorly circumscribed mass composed of lobules and cords of polyhedral to epitheloid cells having centrally located vesicular, round nuclei and abundant granular eosinophilic cytoplasm (Fig 3).No areas of increased abnormal mitotic figures,prominent nucleoli or necrosis were seen.No areas of infiltration at the edges or destructive activity of surrounding muscle fibres was seen,ruling out its malignant nature. Tumour cells exhibited strong and diffuse positivity for immunostain S-100 and positivity for intracytoplasmic granules with periodic acid-Schiff (PAS) staining with resistant to diastase (Fig 4). Some schwannomas and neurofibromas may show granular changes in parts, but the changes are never extensive enough to create a major diagnostic challenge. Moreover, schwannomas are encapsulated which shows central hypointense and peripheral hyperintense (rim sign) on T2 MRI images, these features and other stigmata of von Recklinghausen disease associated with neurofibromas were absent in this case. Rhabdomyomas and hibernomas can show superficial resemblance and may be considered as differentials at deltoid muscle site, however tumour cells do not showed cytoplasmic striations or vacuoles and were negative for skeletal muscle markers and fat stains. Alveolar soft part sarcoma also shows organoid growth pattern and periodic acid-Schiff (PAS)–positive intracellular crystalloids. The characteristic rhomboid crystalloids seen in alveolar soft part sarcoma, were absent in this tumors. Hence, the diagnosis of benign granular cell tumour at unusal site of deltoid muscle was made.

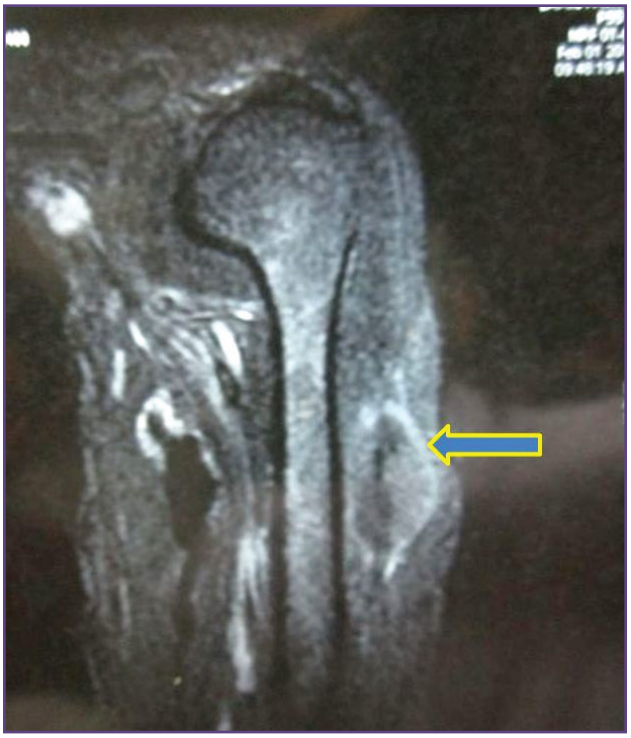

Fig. 1: M.R.I.Scan showing a solitary poorly circumscribed mass, hypointense to muscle on unenhanced T1-weighted images, measuring 5x3x2 cm within deltoid muscle at its distal end.

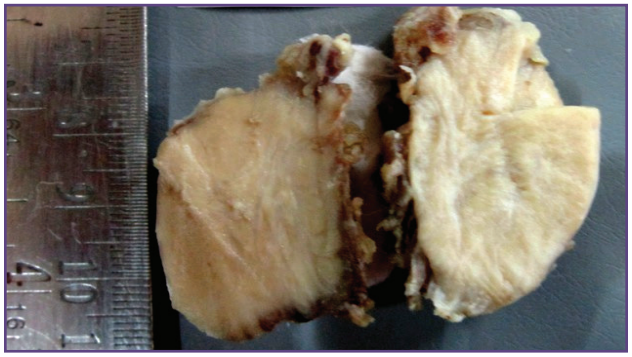

Fig. 2: Gross: Mass measuring 5x3x2 cm, poorly circumscribed, lobulated,homogenous, greyish tan.

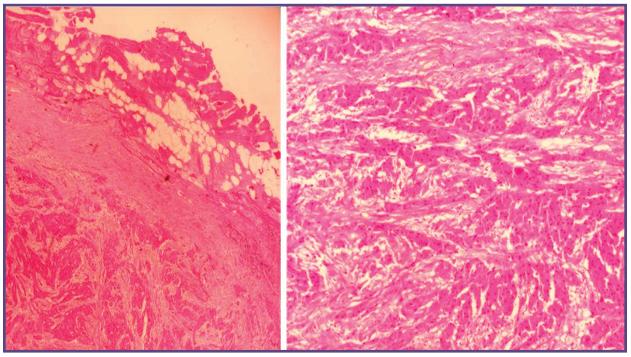

Fig. 3 A: Tumour composed of lobules and cords of polyhedral to epitheloid cells (H & E, x40, x100X)

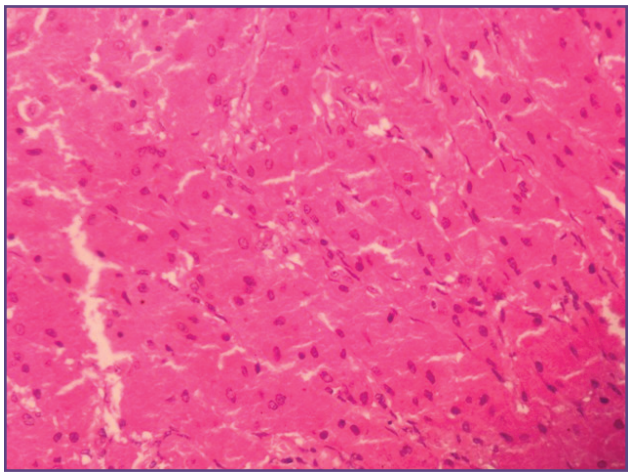

Fig. 3: B: Tumour cells showing centrally located vesicular, round nuclei and abundant granular eosinophilic cytoplasm. (H & E x400X)

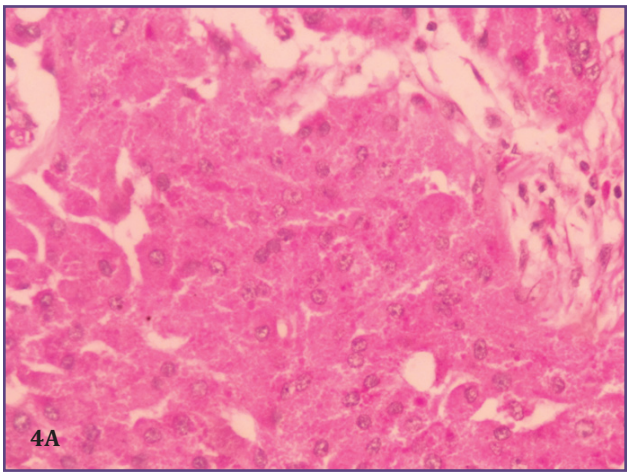

Fig. 4A: Tumour cells showing PAS positive granules in cytoplasm (PAS, x400X),

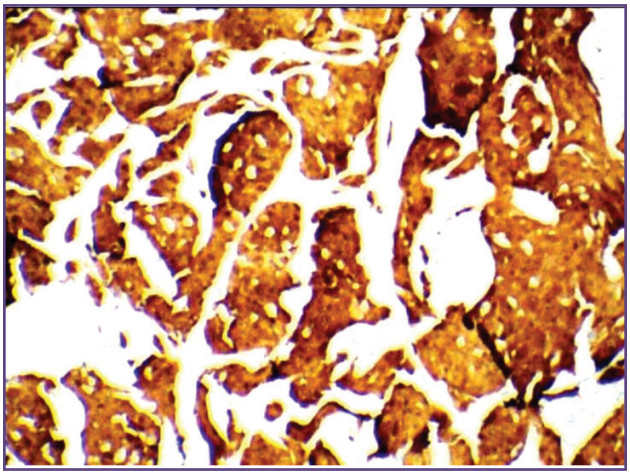

Fig. 4B: Tumour cells showing strong and diffuse immunoreactivity for S-100 immunostain.

Discussion

Granular cell tumours (GCT) are rare and the literature relating to them is sparse. Abrikossoff was the first to describe this tumour in the literature in 1926 [1] as a myoblastoma, since it was reported as a tumour arising from muscle in the tongue. Over the following 60 years the tumour was thought to be neural in origin since it was observed that tumours arising in the extremities were related to the radial and sciatic nerve trunks [2, 3]. Current opinion concurs with this. Although the etiology of GCT is still controversial, the currently most accepted hypothesis is that the tumour arises from Schwann cells or their precursors [3, 4]. Immunohistochemical analysis has shown a strong and consistent positivity for protein S-100, a finding supporting the hypothesis that GCT is of peripheral nerve sheath origin [4].

GCT is a rare tumour that can affect various regions of the body, such as the skin, soft tissues, breast, and lungs [4]. However, GCT is more frequently found in the head and neck region, which accounts for 45% to 65% of all sites affected by the tumour. Of these, 70% are located in the oral cavity, especially the tongue, oral mucosa, and hard palate [4, 5]. Of the reported cases, approximately 30% arise in the oral cavity, with the tongue being the predominant site of occurrence. Another 30% originate in the skin and subcutaneous tissues, while 15% are localised in the breast and 10% in the respiratory tract. The tumour has also been described in skeletal muscles, gallbladder, urinary bladder, female genitals, and the peripheral and central nervous system. [5] The average age of patients is 32 years, with a typical age range of 15 to 60 years. GCT seems to be more prevalent among women, but a gender preference is not unanimously accepted. Tsuchida et al. reviewed the reported cases of granular cell tumours in the English and Japanese literature [6].They found a 2.9: 1 female to male preponderance for the tumour and compared it with previous reported series (1.8:1) and average age 32 years [7]. 26.8% of cases were in the lower extremities and previous reports showed that benign tumours were uncommon (6.4%) in the lower extremities. Of 11 lesions presenting in the upper and lower extremities 36% were within muscle and only 4% of those were benign.

Clinically, benign GCT manifests as a nodular lesion that is generally asymptomatic and solitary, although cases of multiple lesions have been reported [7, 8]. Grossly, granular cell tumours appear as poorly demarcated homogenous, greyish-white or tan nodules, that rarely exceeds 3 cm in diameter. 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. Growth tends to be very slow except for the rarer malignant granular cell tumours, in which growth is generally much more rapid [9].Conventional granular cell tumours are benign neoplasm. Malignancy occurs in less than 2% of patients. 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. In some cases, the epithelium that covers the tumour exhibits pseudoepitheliomatous hyperplasia [7,8,9]. Our case illustrates a benign granular cell tumor in a distinctly unusual location within the deltoid muscle of the arm. Diagnosis was confirmed as benign granular tumour on characteristic histopathological morphology, PAS positive with diastase resistant cytoplasmic granules,intense immunoreactivity for S-100 protein on IHC examination and with clinic-radiological features.

The tumours are typically poorly circumscribed, with the granular cells trailing off into surrounding tissues from the main mass of cells. Although GCT is an uncommon benign neoplasm,cases of malignant GCT have been reported in the literature. Malignancy is diagnosed by a combination of histological findings, including cellular pleomorphism and elevated mitotic activity, and clinical malignant behaviour [9] Clinical suspicion of malignancy should be higher if the lesion is large or growing rapidly, or if there is evidence of distant spread. 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. Fanburg-Smith et al. defined the diagnostic criteria for malignant granular cell tumours and correlated their findings with previous series [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. 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.

Surgical excision with a wide margin is the treatment of choice for Granular cell tumour, although this is not always possible because the tumour lacks a capsule, a condition histologically demonstrated by an undefined cell margin. 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

Conclusion

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. Frozen section was necessary to make the diagnosis, as the imaging findings of granular cell tumour are often non-specific. Pathologists evaluating such mass at unusual deltoid muscle site need to be aware of the entity and give diagnosis with help of imaging, histology, special stains and IHC as it generally carries a good prognosis.

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Dr. A.D. Kalgutkar, Prof & Head, Dept of Pathology, LTMMC & LTMGH, Sion, Mumbai, India

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Competing Interests

None declared

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