CASE REPORT

A rare pediatric cutaneous granular cell tumour of the trunk

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ABSTRACT

Granular cell tumour (GCT) is another rare benign tumour of Schwann cell origin that may arise in virtually any location of the body. We present the case report of a seven-year-old male who presented with a slow-growing, asymptomatic, brownish plaque on the upper right back. Histopathology showed dermal cell proliferation, characterized by numerous eosinophilic granular cytoplasmic inclusions within a sclerotic background. A positive immunohistochemical staining for S-100 and CD68 confirmed the diagnosis of a benign granular cell tumour. The lesion was not very large and remained stable; therefore, the family decided not to excise it immediately, but rather to observe it. This case has highlighted the importance of confirmation through histopathology and immunohistochemistry in suspected GCT, particularly in the pediatric population, where clinical imitation is common. It also emphasises the fact that a conservative approach to management is suitable in cases where the lesion is benign and asymptomatic. (www.actabiomedica.it)

Key words: granular cell tumour, benign, malignant, conservative management.

Introduction

Granular cell tumours (GCTs) are rare neoplasms that were historically referred to as granular cell myoblastomas (1). First explained by Abrikossoff in 1926, these tumours account for roughly 0.5 % of all soft-tissue neoplasms (2). They are usually benign, with fewer than 1–2 % exhibiting malignant behaviour (3). GCTs can occur at any age but are most frequent in adults between the fourth and sixth decades and are reported to be more common in women (4). GCTs may originate in any body parts, most frequently in the tongue (30%), head and neck (70%), breasts (5-15%), and limbs (5). Although GCTs can present at any age, occurrence in

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children is rare. A 2016 Indian case series noted that most GCTs occur between 30 and 60 years of age (6), and reports of paediatric cases are limited (1). Isolated lesions in children may be easily mistaken for benign dermal cysts or appendageal tumours; the unusual location on the trunk can further obscure the diagnosis (1, 4). Recent paediatric case reports describe solitary nodules on the back or finger, emphasising that paediatric GCTs can mimic epidermal cysts or warts and highlighting the need for histopathological assessment to exclude malignant transformation or genetic syndromes (1, 7). The scarcity of paediatric cases underscores the importance of documenting unusual presentations to inform clinicians and avoid delay in management. This case report aims to describe an uncommon paediatric cutaneous granular cell tumour, emphasise the diagnostic value of histopathology and immunohistochemistry in differentiating it from clinical mimics, and justify conservative management with structured surveillance in a child to minimise morbidity while ensuring early detection of recurrence or malignant change risk.

Case presentation

A seven-year-old boy reported to the dermatology clinic with a lesion on the right upper back. The lesion had been present for almost two years, gradually increasing in size over time. The primary parental worries were cosmetic appearance and potential malignancy. The child was else healthy, with normal growth and developmental milestones. There were no associated systemic symptoms such as fever, weight loss, fatigue, or malaise. The child had no known allergies, was not on regular medications, and had no history of previous surgeries or chronic illnesses. Birth history was significant only for low birth weight secondary to maternal gestational diabetes. Family history was unremarkable for neurocutaneous syndromes, hereditary skin disorders, or malignancies. The child had normal school attendance, physical activity, and behaviour. The lesion initially appeared as a small skin-coloured papule and enlarged gradually to approximately 1 × 3 cm over the course of two years.

The lesion was not associated with pain, pruritus, bleeding, ulceration, or discharge. It has not changed in colour, nor has it been seen to become lighter or darker in recent months. It is currently light brown (Figure 1). The lesion has not had any periods of inflammation or infection. There are no similar lesions elsewhere on the body. The patient's appetite, general health, and activity levels are normal. Upon examination, a single, well-defined, firm, brownish plaque was noted on the right upper back, overlying the scapular area. It measured approximately 1 × 2 × 1 cm, with a smooth surface and mild elevation above the level of the surrounding skin. The plaque was mobile over the underlying superficial tissues but not over deeper-seated structures, and there was no associated tenderness, ulceration, scaling, crusting, bleeding, or surrounding erythema. No palpable cervical, axillary, or supraclavicular lymphadenopathy was noted. The oral mucosa was intact, and other mucosal surfaces were unremarkable. The cardiovascular, respiratory, abdominal, and neurologic examinations were normal. The lack of systemic findings and the localised lesion supported a benign aetiology.

Differential diagnosis

Based on the clinical features, the differential diagnosis included:

- Dermatofibroma
- Juvenile xanthogranuloma
- Adnexal neoplasm
- Benign neural tumor (e.g., neurofibroma)

The slow growth, solitary nature, and lack of associated pain or erythema were reassuring signs of a

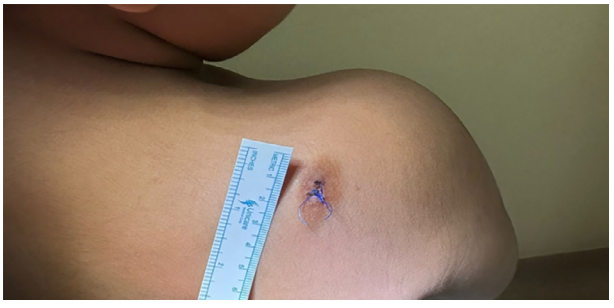

Figure 1. Skin coloured papule on the right upper back.

benign lesion. However, given the unusual presentation, histological examination was recommended to provide a definitive diagnosis and to rule out the rare possibility of malignancy.

Investigations

Histopathology of the punch biopsy showed a well-defined, non-encapsulated dermal lesion, composed of large polygonal cells with coarse eosinophilic granular cytoplasm and small central nuclei (Figure 2). The cells were organised in sheets and nests separated by fine fibrous septa. No necrosis, mitotic activity, or nuclear pleomorphism was observed.

Immunohistochemistry demonstrated

S-100 positivity, confirming Schwann cell origin. CD68 positivity, indicating lysosomal cytoplasmic granularity (Figure 3). These features were coherent with a benign granular cell tumour. Routine laboratory investigations were within normal limits. No imaging studies were required due to lack of deep tissue involvement or systemic disease.

Diagnosis

Benign cutaneous GCT of the right upper back.

Management and follow-Up

Considering the small size of the lesion, the benign histopathology and lack of functional or cosmetic sequelae, and in accordance with family preference following thorough discussion, conservative management was advised. The family was counselled about the very low risk of malignant transformation and reassured that if the lesion changed, complete surgical excision would be curative. The family agreed to a follow-up visit every six months for the first two years to assess changes in size, colour, surface contour, or the development of symptoms. The lesion remained unchanged, with no evidence of recurrence, pain, ulceration, or other changes at the six-month follow-up, and the child continued to be well.

Discussion

GCTs are rare and account for only about 0.5 % of all soft tissue tumours (8). They usually present as solitary, slow-growing, painless nodules and occur most commonly in adults between the fourth and sixth decades of life, with a female predominance and a predilection for head and neck sites such as the tongue (4). Paediatric GCTs are exceptionally uncommon; in a review of paediatric cases, GCTs were noted to be

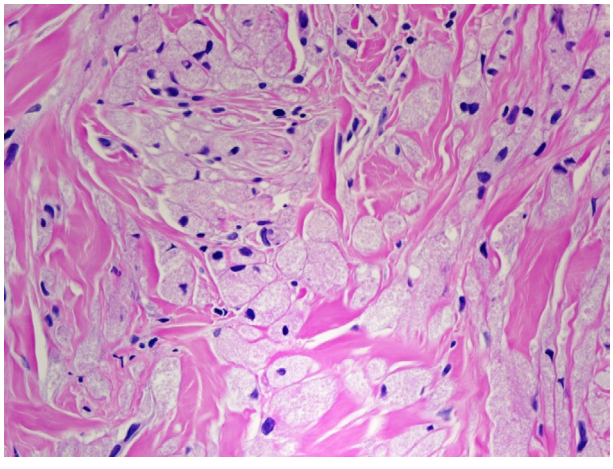

Figure 2. High power view demonstrating large polygonal tumour cells with abundant granular eosinophilic cytoplasm and small uniform nuclei, characteristic of granular cell tumour.

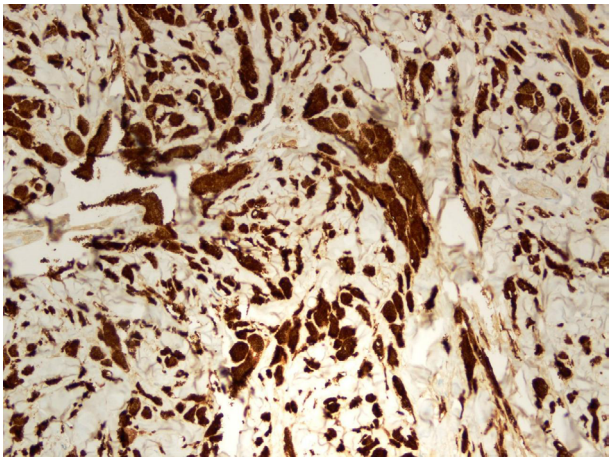

Figure 3. Immunohistochemical stain for S100 shows strong diffuse cytoplasmic and nuclear positivity in the tumour cells, confirming neural differentiation coherent with a GCT.

very rare in children compared with adults (6). When they do occur, lesions may be mistaken for more common benign dermal papules such as dermatofibromas or neurofibromas (4). The case under discussion involved a solitary, slowly enlarging cutaneous lesion on the trunk of a 7-year-old child. This is a particularly atypical presentation because the most of reported GCTs arise in the oral cavity or head and neck, and paediatric cutaneous lesions on the trunk have been reported only sporadically (4). Awareness of this rare age group and anatomical presentation is therefore essential to prevent misdiagnosis and to prompt appropriate investigation. Clinically, GCTs have non-specific appearances and often resemble other benign lesions; therefore, histopathological examination is essential for diagnosis. On haematoxylin–eosin staining, GCTs are characterized by sheets of polygonal cells infiltrating the dermis or submucosa. These cells have abundant eosinophilic granular cytoplasm and small centrally placed nuclei (4). The granularity results from numerous lysosomes, and the granules are periodic acid–Schiff positive and diastase resistant (4). Immunohistochemistry further supports the diagnosis: tumour cells typically express S-100 protein, neuron-specific enolase, CD68 and other neural markers, with negativity for cytokeratin and desmin (4). In a cutaneous GCT case on the trunk, neoplastic cells demonstrated centrally located nuclei with granular eosinophilic cytoplasm and stained positively for S100, neuron-specific enolase and CD68 (8). These markers confirm Schwann-cell origin and help to differentiate GCTs from histologic mimics such as rhabdomyoma, juvenile xanthogranuloma and adnexal tumours. Assessment of malignancy relies on histologic criteria. Malignant transformation is rare (<2 % of cases) (9), but malignant GCTs tend to be larger (>4 cm), show rapid growth, ulceration, or metastatic spread. Histopathologic features suggestive of malignancy include necrosis, nuclear pleomorphism, spindling, increased mitotic activity, high nuclear-to-cytoplasmic ratio and vesicular nuclei with prominent nucleoli (4). Fanburg-Smith *et al.* (1998) (10) proposed six criteria for a histopathological diagnosis of malignancy; these include spindling, necrosis, increased mitotic activity, vesicular nuclei with large nucleoli, pleomorphism, and high nuclear to cytoplasmic ratio. The presence of ≥ 3 features fulfil the Fanburg-Smith criteria for malignancy (10). In the current case the lesion was small, lacked atypia, and showed none of these malignant features on microscopy, supporting a benign diagnosis. Once a GCT is confirmed as benign, management must balance complete excision against the risks of surgery, particularly in children. Traditional management advocates complete surgical excision with clear margins to minimise recurrence (11, 12). However, recurrence rates of 2–8 % occur even when margins are histologically clear, and rates can reach 20 % when margins are positive. Malignant transformation remains very rare (1–2 %) (13). Because of this generally indolent behaviour, some authors propose a conservative approach for small lesions. Ferraz (2021) notes that a conservative approach with regular follow-up is appropriate for tumours <10 mm in diameter without features of malignancy, whereas surgical excision is reserved for lesions >20 mm, symptomatic lesions or tumours with suspected malignancy (9). This guidance supports watchful waiting in carefully selected cases, particularly when the lesion is small, asymptomatic and cosmetically acceptable, as in the current patient. In paediatric patients, avoiding surgery can be desirable to spare general anaesthesia and scarring. Families should be counselled that benign GCTs rarely metastasise but can recur if incompletely excised (13). They should also be educated on features suggestive of malignant change, such as rapid growth, ulceration or lymphadenopathy. Close clinical surveillance with periodic review (e.g., every six months) is advisable; imaging or repeat biopsy should be considered if the lesion enlarges or changes. In case of recurrent growth or if cosmetic concerns arise, surgical excision remains curative. In our case, the lesion's small size and benign histology rationalised a conservative approach with six-monthly follow-up, recognising that surgery could be offered later if needed.

Conclusion

Granular cell tumours are rare in children, may present at atypical sites on the trunk and other sites and may be easily misdiagnosed. Recognition of their clinical spectrum, along with early biopsy, is key.

Histopathology and immunohistochemistry will remain key to diagnosis and to differentiate benign and malignant lesions. Small asymptomatic lesions without malignant characteristics, which mainly occur in children, should be managed conservatively, with regular surveillance.

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