Granular cell tumor in a child: An uncommon cutaneous presentation

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ABSTRACT

Granular cell tumors (GCTs) are uncommon soft tissue tumors which present as papulonodular lesions and are often diagnosed histopathologically. These usually develop in adulthood and are considered to be of Schwann cell origin. Most of the lesions are benign, but malignant lesions with poor prognosis are known to occur. We report a case of GCT in a 9-year-old girl presenting as an isolated lesion simulating an acrochordon. The histopathological and immunohistochemical evaluation showing polygonal granular cells positive for S-100 and neuron-specific enolase, and negative for cytokeratin and desmin helped clinch the diagnosis. Complete excision under local anesthesia was done. The atypical clinical morphology and diagnostic histopathology of this uncommon entity are presented to aid the clinician in recognizing it. These tumors are mostly benign, rarely malignant, with the latter category having a poor prognosis. A near-complete excision is recommended in view of the minimal risk of malignant transformation.

Key words: Atypical presentation, child, granular cell tumor, polypoidal

Access this article online

Website: www.idoj.in

DOI: 10.4103/2229-5178.190494

Quick Response Code:

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INTRODUCTION

Granular cell tumor (GCT) is an uncommon soft tissue tumor with a peak incidence in the fourth–sixth decade. It is more common in women and people of African descent. Lesions involve skin and soft tissue, even the viscera, with most of these arising in head and neck area. Tongue is the most common site of origin.[1] Cutaneous lesions present with a nonspecific papulonodular morphology, and histopathology is required for a precise diagnosis.

We present a case of an asymptomatic, polypoidal swelling arising over the buttock in a 9 year-old-girl, which was diagnosed as a GCT. The atypical clinical presentation, in a young child with the typical histopathology, is discussed.

CASE REPORT

A 9-year-old girl presented with a single, asymptomatic, raised lesion over the right buttock for one and a half month. On examination, a solitary 1.5 cm sized, soft, nontender, exophytic growth with grape-like surface was seen on the right buttock [Figure 1]. There was no mucosal involvement, regional lymphadenopathy, history of rapid growth, or ulceration. The child was developmentally normal, attending school regularly. An initial differential diagnosis of acrochordon, solitary neurofibroma, premalignant fibroepithelial tumor of Pinkus, or a benign appendageal tumor was entertained. The lesion was excised and sent for histopathology.

Hematoxylin and eosin stained sections revealed a polypoidal morphology with mild hyperkeratosis and acanthosis [Figure 2a]. Sheets of polygonal cells with abundant eosinophilic granular cytoplasm diffusely infiltrating the papillary and reticular dermis were seen [Figure 2b]. Most of these had a centrally placed nucleus and a single conspicuous nucleolus; while in a few, it was placed eccentrically.

Cite this article as: Daulatabad D, Grover C, Tanveer N, Bansal D. Granular cell tumor in a child: An uncommon cutaneous presentation. Indian Dermatol Online J 2016;7:390-2.

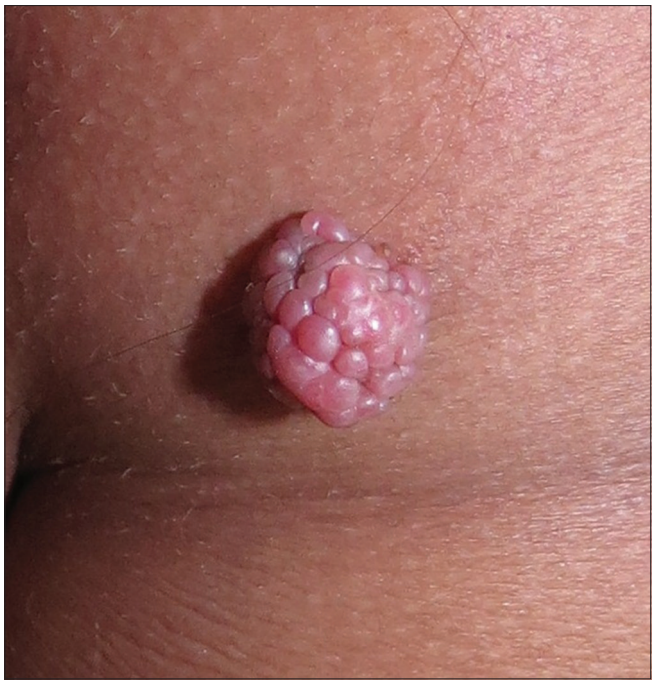

Figure 1: A single 1.5 cm sized pedunculated grape-like growth on the right buttock of a 9-year-old girl

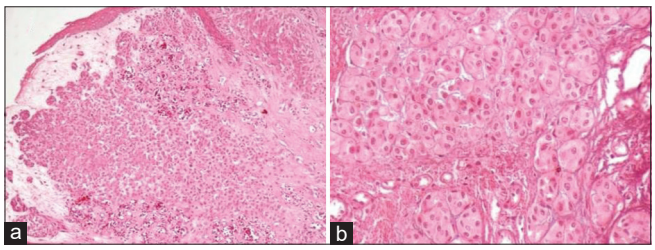

Figure 2: Hematoxylin and eosin stained sections of the excised tissue shows (a) a polypoidal growth with hyperkeratosis of the epidermis and diffusely infiltrating sheets of polygonal cells in the papillary and reticular dermis (100x). (b) High power view showing abundant eosinophilic granular cytoplasm (400x)

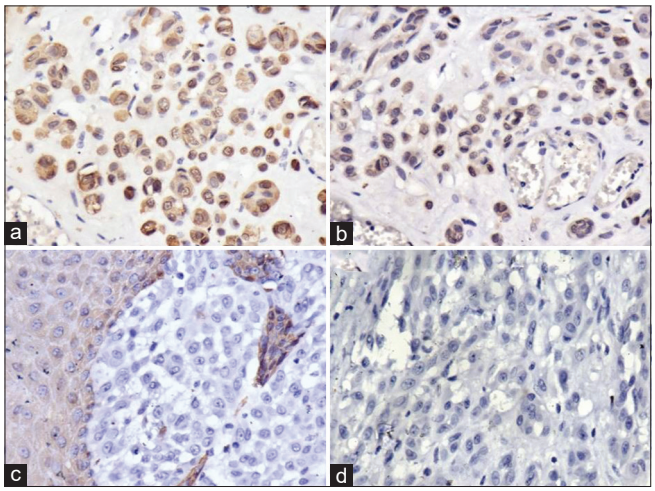

Figure 3: Immunohistochemistry revealed positivity to S-100 (a) and neuron specific enolase (b) and lack of staining for cytokeratin (c) and desmin (d) as seen on × 400

There was an absence of pleomorphism, necrosis, or mitosis. Immunohistochemical staining revealed the tumor cells to be positive for S-100 [Figure 3a] and neuron-specific enolase (NSE) [Figure 3b] and negative for cytokeratin [Figure 3c] and desmin [Figure 3d]. A diagnosis of benign GCT was entertained. On further 6 months follow-up, there was no local recurrence or any new lesions.

DISCUSSION

GCT was described by Weber in 1854. In 1926, Abrikossof established it as a separate histopathological entity. It is also known as Abrikossof's tumor or granular cell myoblastoma.[1] The origin of GCT has long been a topic of dispute; however, electron microscopic findings reveal the presence of axonal structures and immunohistochemical studies demonstrate S-100 positivity, suggesting a Schwann cell origin.[2]

Clinically, the tumor develops in adults as a solitary, asymptomatic, dermal or subcutaneous papule or nodule, which is generally benign. It is often misdiagnosed as a dermatofibroma, appendageal tumor, neurofibroma, or angiolipoma. Presentation as an exophytic lesion in a child, as was seen in our patient, is an unusual morphology. Other reports of atypical presentation include a 43-year-old Japanese male presenting with a papillomatous but sessile lesion,[3] and a 53-year-old woman presenting as hyperpigmented panniculitis-like lesion on the arm.[4] Multiple GCTs have been reported rarely, especially in children, in association with neurofibromatosis, Noonan's syndrome, Watson's syndrome, or growth retardation.[5]

The diagnosis is often clinched on histopathology. The tumor cells have abundant granular eosinophilic cytoplasm with centrally located vesicular or pyknotic nuclei. Granular appearance is due to the presence of large lysosomes, staining positive with periodic acid-Schiff (PAS) stain, and resistant to diastase. They can also be stained with Sudan black B and trichrome preparations. Immunohistochemistry reveals expression of S-100, CD68, NSE, nerve growth factor receptor 5, microphthalmia-associated transcription factor, inhibin alpha, and protein gene product 9.5.[6] These cells may be arranged in sheets or nests with variable stroma. In the present case, we considered rhabdomyoma, GCT, and rhabdoid tumor initially, due to an abundance of cytoplasm and few cells with eccentric nuclei; however, immunohistochemistry firmly established the diagnosis.

Malignant transformation is extremely rare (<2% cases). Clinically, such lesions may present with size >4 cm, lymph node metastases, aggressive clinical behavior, rapid growth, and ulceration. Suggestive histopathological features include

nuclear pleomorphism, increased mitotic activity, and necrosis. Fanburg-Smith *et al*. proposed six criteria for a histopathological diagnosis of malignancy; these include necrosis, spindling, vesicular nuclei with large nucleoli, increased mitotic activity, high nuclear to cytoplasmic ratio, and pleomorphism. The presence of three or more of these qualifies as a malignant tumor.[7]

The treatment of choice is complete surgical excision. Histopathology should rule out the signs of malignant change, although it is rare. Excision was successful in our patient with no recurrence even after 6 months of follow-up.

The present case is unique because of distinctive morphology in the form of an exophytic, soft growth with a polypoidal appearance (simulating a bunch of grapes) arising in a young child. To the best of our knowledge, such a presentation has only been reported once.[3] The case serves to highlight that complete excision and histopathological evaluation should be attempted even in mildly suspicious cases to offer a complete cure.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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