Multiple granular cell tumours in a 10 year old boy – a case report

T D P Lokunarangoda[1], J Femando[2], S P W Kumarasinghe[3]

Sri Lanka Journal of Dermatology, 2005, 9, 41-42

Granular cell tumour is an uncommon benign tumour, usually occurring in the skin as a single nodule[1]. The first case of granular cell tumour was described by Weber in 1854. Subsequently Abrikossoff established this tumour as a clinical entity in 1926[1].

Multiple granular cell tumours are very rare, particularly in children. Only a few reports have described the occurrence of multiple granular cell tumours. Here we describe multiple granular cell tumours of the skin in a 10 year old boy.

Case report

A 10 year old boy developed multiple nodules on the posterior neck, left shoulder, right wrist and right foot. These nodules were asymptomatic and slow growing over two years. There were no other systemic symptoms. Development of the child had been normal. Family members had no similar lesions.

Dermatological examination revealed four; firm, nontender nodules on posterior neck, left shoulder, right wrist and right foot. The size of the lesion varied from 5 to 7mm. Overlying skin was normal. No other dermatological neurological or develop-mental abnormalities were noted. Clinical differential diagnoses considered were neurofibroma, dermatofibroma, pilomatricoma, and steatocystoma multiplex.

Two excision biopsies of the posterior neck lesions and a right shoulder lesion were done. Histological examination of both tumours revealed focal areas of large cells with centrally placed nuclei and granular eosonophilic cytoplasm in the dermis. Overlying skin was normal. There was no evidence of nuclear atypia or mitoses. A diagnosis of granular cell tumour was made.

Histology of the biopsies was critically evaluated in the light of clinical differential diagnoses. Histology of dermatofibroma usually has two cell types which consists of small compactly arranged fibroblasts with spindle shaped nuclei and scanty cytoplasm and histiocytes which usually have prominent pale staining cytoplasm full of lipid[2]. Pilomatricoma has irregular islands of dark staining basaloid cells which become eosinophilic and have a central zone representing the lost nucleus [termed shallow cells]. Calcification and a brisk foreign body giant cell reaction to the hair matrix material frequently dominates the histology[3]. Steatocystomas are thin walled cysts. The main diagnostic feature is a thin brightly eosonophilic almost glassy band of keratin lining the cyst with underlying, flattened squamous cells[3]. Neurofibroma is composed of spindle shaped cells embedded in wavy reticulin and collagen fibers[2]. In our case focal areas of large cells with centrally placed nuclei and granular eosonophilic cytoplasm in the dermis. These granules were PAS positive which confirmed the diagnosis of granular cell tumour.

Discussion

Granular cell tumour generally occurs as a small poorly circumscribed nodule that may be solitary or multiple and pursues a benign clinical course. It occurs in patients of any age, but is commoner in 4h to 6h decades of life and is rare in children. Granular cell tumours most frequently occur as solitary lesions on the tongue and skin of middle aged persons[4]. Skin and subcutaneous tissue and mucosae of the head and neck are areas of predilection for granular cell tumour[5].

Exact nature of granular cell tumour is still unclear with conflicting evidence for neural, pericytic and smooth muscle origin[4]. Most of cutaneous and subcutaneous granular cell tumours are currently thought to arise from the Schwann-cell[6] neurological origin. The finding that granular cell tumours react with S 100 antibodies further supports it. A case report provides evidence for an association between multiple granular cell tumours in childhood and neurofibromatosis[4]. Granular cell tumour is an

[1]*Senior Registrar in Dermatology*, [2]*Consultant Pathologist*, [3]*Consultant Dermatologist.*

uncommon tumor found on the hands and feet. It may be associated with a peripheral nerve. Its presentation in the fingers or hand is often painful[9]. It has been claimed that this is a characteristic that is not well recognized. However in our patient the lesions were neither painful nor tender.

Selter et al has reported multiple granular cell tumours and growth hormone deficiency in a child[1]. Multiple granular cell tumours in a child's finger has been reported by Price et al[7]. The congenital epulls or congenital granular cell tumour is a rare gingival tumour occurring in infancy that often regresses spontaneously[4]. Most of reports on the cytologic features of granular cell tumours have been on the lesions from breast and respiratory tract.

A malignant granular cell tumour is recognized but requires rigorous histological confirmation. Malignant granular cell tumours have not been reported in childhood and adolescence[4].

An association with multiple granular tumours and Phakomatosis pigmentovascularis has been reported[9,10]. Most reported cases of Phakomatosis pigmentovascularis have concerned Asians. There are pigmentary as well as associated vascular anormalies. Dermatological features include multiple Mongolian spots, naevus of Ota, Port wine stain, naevus spilus and naevus anaemicus. Out of four major types of Phakomatosis pigmentovascularis, type III is associated with multiple granular tumours[9,10].

Our case is uncommon because the lesions were multiple without any other associations and he is a child of 10 yrs. Furthermore lesions on the foot have not been discribed before.

References

1. Seiter S, Ugurel S, Tilgen W, Reinhold U. Multiple granular cell tumors and growth hormone deficiency in a child. *Pediatr Dermatol.* 1999; **16**(4): 308-10.
2. Epidermal skin appendage harmatomas and tumours, In: Stevans A, Wheater P. R., Lowe J. S, editors. Clinical Dermatopathology. London: Churchill Livingstone, 1989: 155-57.
3. Fibrous and fibrocystic tumours and proliferation, In: Stevans A, Wheater P. R., Lowe J.S, editors. Clinical Dermatopathology. London: Churchill Livingstone, 1989: 189,183.
4. Burgdorf WHC, Ruiz-Maldonado R. Benign and malignant tumours. In: Schacher L. A, Hansen R.C, editors. Paediatric Dermatology. London: Mosby, 2003: 876.
5. Kershisnik M, Batsakis JG, Mackay B. Granular cell tumors. *Ann Otol Rhinol Laryngol.* 1994 ; **103**: 416-9.
6. LeBoit PE, Barr RJ, Burall S, Metcalf JS, Yen TS, Wick MR. Primitive polypold granular-cell tumor and other cutaneous granular-cell neoplasms of apparent nonneural origin. *Am J Surg Pathol.* 1991; **15**(1): 48-58.
7. Price W, Paletta CE, Woodberry KM. Granular cell tumor in a child's finger. *Ann Plast Surg.* 2000 ; **44**(4): 447-50.
8. Mallik MK, Das DK, Francis IM, al-Abdulghani R, Pathan SK, Sheikh ZA, Luthra UK. Fine needle aspiration cytology diagnosis of a cutaneous granular cell tumor in a 7-year-old child. *Acta Cytol.* 2001; **45**(2): 263-6.
9. Kumarasighe. P, Chio MTW. Pigmentary disorders. In: Goh. C. L, Chua. S. H, NG. S. K, editors. The Asian skin. Singapore: McGraw Hill, 2005: 59.
10. Guilia K C, Prendiville JS. Multiple granular cell tumours associated with giant speckled lentigenous naevus and naevus flammeus in a child. *J Am Acad Deratol.* 1999; **24**: 359-63.