Giant gluteal lipoblastoma associated with hepatic haemangioma and bilateral nephromegaly

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DESCRIPTION

An 11-month-old girl adopted from Guatemala was referred to us with a painless static massive swelling in the right buttock of 6 months duration. It was soft, mobile, lobulated and measured 6×8 cm. Ultrasound demonstrated a heterogeneous echogenic mass with a little Doppler flow within it. There was a small focal hypervascular lesion in the right lobe of the liver suggestive of a haemangioma. Both the kidneys were enlarged but normal. CT scan confirmed hepatic haemangioma and bilateral nephromegaly (figure 1A). It showed a large soft tissue mass of background fatty attenuation with abundant strands of enhancing soft tissue in the right gluteal region suggesting lipoblastoma (figures 1B–D). An MRI scan showed a well-defined predominately fatty signal of diameter 7 cm in the right gluteal muscles and confirmed hepatic haemangioma and bilateral nephromegaly. Biopsy confirmed lipoblastoma. She underwent total excision of lipoblastoma uneventfully. Histology showed lobulated architecture, with lobules of lipocytes and lipoblasts separated by prominent fibrovascular septae suggestive of lipoblastoma. She has been followed with serial ultrasound scans and

the hepatic haemangioma slowly regressed and spontaneously resolved at 21 months, nephromegaly resolved to 50th centile at the age of 4 years and there is no tumour recurrence at 6 years follow-up. Lipoblastoma is a rare benign lipomatous tumour arising from embryonic white fat and encountered almost exclusively in infants and young children.¹ Surgical excision is the treatment of choice but spontaneous resolution has been reported.² In infancy and early childhood, the identification of a tumour composed mostly of fat should suggest the diagnosis of lipoblastoma or lipoblastomatosis.³ The coincidence of nephromegaly and liver haemangioendotheliomas has been reported in Beckwith-Wiedemann syndrome and other associated syndromes.^{4 5} Nephromegaly means enlargement of kidneys which could be unilateral or bilateral and congenital or acquired. Congenital bilateral nephromegaly in association with live haemangioma and lipoblastoma may represent a variant of Beckwith-Wiedemann syndrome. It may represent nephroblastomatosis or it may regress as the patient grows older. In isolated nephromegaly, it may increase the risk of renal scarring if associated with vesicoureteral reflux.⁶

Figure 1 CT scan showing (A) hepatic haemangioma and bilateral nephromegaly (RK, LK), (B–D)—top, middle and bottom cuts of the right gluteal tumour with dumbbell extension into the intermuscular plane.

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Learning points

- ▶ Lipoblastomas occur on the extremities, common in children below 3 years with a male dominance.
- ▶ A follow-up period of 2–5 years is recommended as the recurrence rate is 14–25%.
- ▶ Lipoblastomatosis is a diffuse form with deeply embedded, non-capsulating, multicentric and infiltrative tumour, and wide local excision is advised.

Competing interests None.

Patient consent Obtained.

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