Solitary mastocytoma causing recurrent blistering in infancy

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Abstract
Two children with solitary mastocytoma in infancy, both of whom had recurrently blistering scalp lesions, are reported in order to emphasise the diagnostic importance of this distinctive history.

Cutaneous mastocytosis in infancy or childhood usually presents with multiple lesions but mastocytomas are solitary in about 10% of cases. Isolated lesions may not be easy to diagnose on clinical appearances but nearly always have a history of recurrent blistering. We report two typical cases of solitary mastocytoma in order to emphasise the importance of this history.

Case reports
CASE 1
An otherwise healthy girl born at term was noted at birth to have a scabbed occipital lesion attributed at the time to a scalp electrode. At 9 days an area adjacent to the scab became swollen and cystic; aspiration produced sterile serosanguinous fluid. The possibility of an occipital meningoencephalocele was considered but excluded by ultrasonography. In the next few weeks the lesion swelled intermittently on five further occasions (fig 1), each time subsiding over a few days during antibiotic treatment. When seen by one of us at 11 weeks the diagnosis of mastocytoma was considered, particularly as the quiescent lesion urticated when rubbed. There was no other evidence of mastocytosis. A small diagnostic biopsy was arranged. At the end of the procedure the surgeon noted that the child went transiently 'bright red from top to toe' without disturbance of vital signs. Histology confirmed mastocytoma (fig 2). Over subsequent months the lesion continued to urticate periodically, and the infant had several episodes of flushing and restlessness each lasting an hour, but unrelated to change in the lesions. By the age of 3 years, all these episodes had ceased, and the lesion had become simply an area of redness with some overlying hair loss.

Histology was that of a mastocytoma, and he was referred to us. When seen, the incompletely excised lesion consisted simply of redness and coarse scale. Two other 0-5 cm telangiectatic macules were present on right thigh and buttock, but none of the lesions urticated on rubbing. Despite the excision biopsy, the scalp lesion has continued to blister intermittently, but there has been no generalised flushing.

CASE 2
An infant boy had a scalp lesion noted at birth which blistered on at least three subsequent occasions. The episodes were treated by aspiration and antibiotics by his general practitioner. At the age of 6 months he was referred to a surgeon who suspected an infected sebaceous cyst and attempted excision of the lesion.

Figure 1 A photograph, taken by the parents, of the lesion during an episode of blistering.

Figure 2 Histology showing mast cells densely packed within the upper dermis.
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Discussion
We wish to draw attention to this disorder, because while relatively uncommon, the presentation is distinctive, and earlier recognition of our cases might have avoided some unnecessary investigation and treatment.

The commonest presentation of cutaneous mastocytosis in childhood is urticaria pigmentosa appearing as numerous small round or ovoid red or red-brown macules or papules, usually in the first six months to two years of life. In most cases the lesions urticate on rubbing, due to mediator release (Darier’s sign), but vesiculation or frank blistering is common in lesions in infancy.

A multiplicity of these nodular or blistering lesions suggests the diagnosis but our two cases suggest that diagnosis of mastocytomas which are solitary is less easy. However, 38 cases of solitary mastocytomas in four early series indicate that the presentation is just as characteristic. Half the solitary lesions were present at birth, most by 3 months, and all but five by the age of 2 years. The usual site was on the trunk or limbs, but not palms or soles; in contrast with our cases, only two of 38 had head or neck lesions. Lesions were yellow, tan or red-brown in colour, slightly elevated or macular, and ovoid, 0-5 to 3 cm in largest diameter. On the basis of these appearances, the clinical differential diagnosis included melanocytic naevi, xanthomas, or juvenile xanthogranulomas. The diagnosis of solitary mastocytoma was suggested by a positive Darier’s sign in only half the cases, but a history of blistering was recorded in 25 and probably more of the cases. Other cases have experienced episodic flushing. The diagnosis can be confirmed by biopsy; unfortunately examination of the serosanguinous blister fluid is not useful as it usually contains neutrophils rather than mast cells. The course of solitary mastocytoma is benign; those lesions not cured by excision appear to improve or resolve during early childhood.

Blistering mastocytomas in the neonatal period or in infancy have initially been diagnosed as bullous impetigo or epidermolysis bullosa, or even cigarette burns (Camille Lazarro, personal communication). It is therefore important for appropriate management to recognise that in infants a history of episodic blistering or swelling at a single site is almost pathognomonic of solitary mastocytoma.

Pituitary gigantism

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Abstract
A case of pituitary gigantism resulting from a pituitary adenoma which secreted growth hormone is described. The patient was successfully treated by surgery, which led to the normalisation of endogenous growth hormone secretion. An acceptable final height was achieved with high dose intramuscular testosterone treatment.

The management of pituitary gigantism is difficult and often requires multiple approaches including surgery, radiotherapy, and medical treatment. It has not been established whether surgery alone can normalise spontaneous growth hormone secretion, for there is the possibility of an underlying abnormality in the secretion of growth hormone releasing hormone and somatostatin. We report our successful experience in the transsphenoidal removal of a pituitary adenoma and the restoration of normal growth hormone secretion in a boy with gigantism.

Case history
A 13 year old boy presented with a five year history of rapid growth. He was born at full term with a weight of 3200 g and length of 51 cm. He grew at a normal rate until 7 years of age when it was noticed he had large hands and required increasing shoe sizes. At the age of 10 years he was taller than a brother 14 months his senior. Immediately before referral he grew 20 cm over a 10 month period. There was no family history of tall stature (mother’s height 163 cm, father’s height 178 cm) and his four siblings were of average height.

On examination his height was 187.5 cm (height SD score +3.57) and his weight was 78 kg. He was in Tanner stage 4 puberty (testes 12 ml). His hands and feet were large with slight sweating. Mild prognathism was noticed. His bone age corresponded to a male standard of 13 years according to the Greulich-Pyle standard. His predicted final height, calculated according to the method of Bailey-Pinneau, was 214 cm.

A cranial computed tomogram demonstrated a pituitary fossa of normal size containing an intrasellar pituitary mass of 12 mm diameter (fig 1).

ENDOCRINE INVESTIGATIONS
Spontaneous nocturnal growth hormone secretion was assessed by taking blood samples at 20
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