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CASE REPORT

Case report of buccal lipoblastomatosis in a 6-month-old infant

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Summary Lipoblastomatosis and lipoblastomas are rare benign tumours arising from fetal—embryonal adipocytes. These tumours occur predominately in young children on their extremities. Chung and Ezinger (E.B. Chung, F.M. Ezinger, Benign lipoblastomatosis. An analysis of 35 cases. Cancer 32 (1973) 82—92) differentiated between the two types and described a lipoblastoma to be localised, and well-circumscribed arising from superficial fat tissue, and a lipoblastamatosis to be an unencapsulated, diffuse-type lesion that grows from deeply situated adipose tissue.

A case of a lipoblastomatosis in a 6-month-old infant is described. There are no reported cases of this tumour occurring in the facial—buccal region. Although these tumours are rare they should be included in differential diagnoses of soft tissue lesions in infants.

Case report

A 6-month-old Caucasian girl was referred to the Oral and Maxillofacial Department at Addenbrooke's Hospital for the assessment of a small lump in the right cheek that had gradually increased in size since the age of 6 weeks. She had initially been assessed by the Consultant Paediatrician at Ipswich Hospital following a referral from her general medical practitioner. An ultrasound scan of the right cheek had been performed and showed an indistinct 'mass' approximately 1 cm in size, lateral to the edge of the lips and was clinically palpable, possibly representing a small lipoma. It had begun to affect the infant's ability to breast-feed on the right side, and the parents were concerned about the facial disfigurement.

Her medical history was unremarkable. She had not developed any primary dentition. Oral examination revealed a 2 cm lump in the right mucosa. It was non-ulcerated, incompressible, non-fluctuant and no reported tenderness. The facial nerve was deemed intact. The case was discussed with senior consultants and an incisional biopsy of the lesion was organised under general anaesthesia.

Treatment

The lesion was fully examined and its approximate dimensions recorded as 3 cm × 2cm × 2 cm. Histopathology of the initial biopsy was reported as con-

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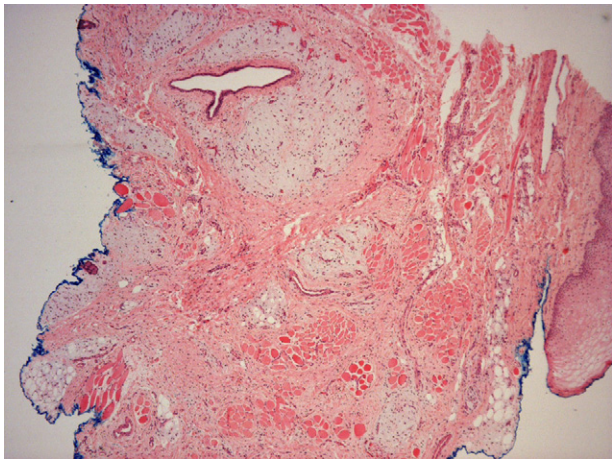

Fig. 1 Specimen showing islands of myxoid grey-white primitive adipose tissue extending through the stroma, with residual islands of skeletal muscle (40x magnification).

sistent with a lipoblastoma. The best management for this was surgical enucleation. This was discussed with the parents. The patient was subsequently admitted for the removal of the benign tumour. A peritumoral incision was made into the mucosa. Blunt dissection was carried out around the mass and bipolar diathermy used as necessary. The specimen was sent for histopathological examination. The post-operative period was unremarkable and the child was discharged the following day.

Histopathological examination revealed a nodular lesion with a diffuse infiltrative margin extending into skeletal muscle (Fig. 1) and mixed salivary tissue. The nodules were composed of lobules of mature and primitive adipose tissue (Fig. 2) with a myxoid appearance in places and delineated by fibrous septa. Numerous lipoblasts were present along with spindle and stellate cells more reminiscent of primitive mesenchyme. There was no significant pleomorphism and mitotic figures were inconspicuous. Overall, the features of primitive adipose tissue and infiltrative margin represent a lipoblastomatosis rather than a lipoblastoma. The lesion extended to margins of resection in several places and therefore complete excision was not achieved. Regular follow-ups of the child would be needed as there is a risk of recurrence.

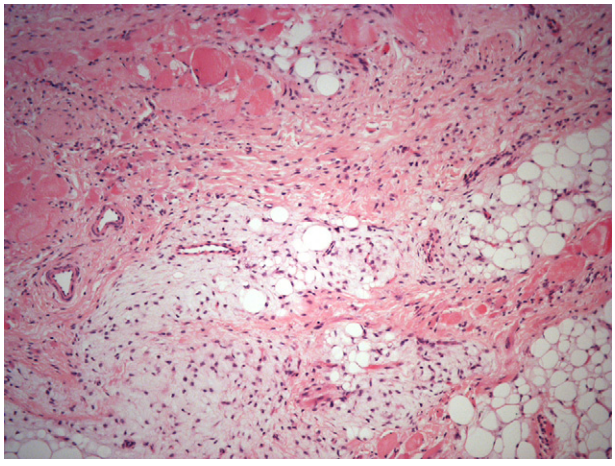

Fig. 2 Specimen showing primitive and mature adipose tissue infiltrating into skeletal muscle (200x magnification).

Discussion

Lipoblastomas are rare benign tumours arising from the fetal—embryonal adipose tissue. In 1926, Jaffe [2] provided the first description of a lipoblastoma when he used the term to describe a lipomatous tumour noted in the groin. The lipoblastoma was comprised of cells resembling embryonic fat cells, which did not metastasise or recur. In 1958, Velios [3,4] proposed another term—lipoblastomatosis, which described a tumour that was on histological examination similar to myxoid liposarcoma however remained circumscribed and showed no signs of invasion to surrounding tissue. He suggested that this tumour developed from postnatal proliferation of lipoblasts. Chung and Ezinger [1] clarified the terminology of these benign tumours, based on their collection of case studies. They stated that a lipoblastoma is a tumour that is localised, and well-circumscribed arising from superficial fat tissue, and lipoblastamatosis is an unencapsulated, diffuse tumour that derives from deeply situated adipose cells. These terms have been universally accepted and used widely over recent years.

A literature review by Kransdorf [6] showed that 114 out of 18,677 soft tissue lesions were lipoblastomas (0.6%), highlighting its rare occurrence. Hicks et al. [9] recorded 26 cases of lipoblastomatous tumours over a 15-year period, of which the majority were lipoblastomas (73%) and the minority lipoblastomatosis (23%).

These tumours predominantly occur in infants and young children under the age of 3 (90%) [5]. However, older patients have been recorded ageing from 5 to 18 years old [1]. Lipoblastomas tend to occur in the superficial subcutis while diffuse lipoblastomatosis is found deep in soft tissues, the mediastinum, retroperitoneum, thorax and abdomen [9]. There have been no reported cases of a lipoblastoma occurring in the buccal area, although other rare sites have included the parotid gland [12], eyelid and tonsillar fossa [11,13]. Case studies by Chung and Ezinger [1] and Collins and Chatten [8]

suggest that lipoblastomatous tumours are most likely to occur on the extremities as noted by 72% of cases. Collins and Chatten [8] noted that 13 out of 18 cases of lipoblastomatous tumours occurred on the left side of the body. The increased incidence of left-sided lesions may suggest a genetic association with genes determining asymmetry.

Lipoblastomatous tumours were more commonly found in males than females in many series, (1.3—3.8 ratio) [2,4,8,7], however in a conflicting report Hicks et al. [9] stated there was no gender predilection. Collins and Chatten [7] reported tumour size ranging from 1.0 to 21.0cm, of which 60% of all tumours were greater than or equal to 5 cm in the greatest dimension. Lipoblastoma are classically rapidly enlarging tumours, usually asymptomatic and presenting as a soft, non-tender mass [1,2,9]. The lesions may present with prominent regularly distributed blood vessels mimicking a haemangioma [3].

Accurate pre-operative diagnosis can be challenging and is reliant on using imaging modalities such as MRI, CT and ultrasound as well as fine needle aspiration cytology or incisional biopsy [14]. Differential diagnoses may include: sarcoma, neuroblastoma, germ cell tumour, lymphoma, epidermal inclusion cyst, lymphangioma, lipoma and Baker's cyst [3,8].

Lipoblastomatous tumours resemble myxoid liposarcomas especially in older children and adolescents. Although this is a rare occurrence it still does occur and should be separated from lipoblastomatous tumours. Shmookler and Ezinger [10,11] described an 8-month-old male with round cell liposarcoma and a 3-year-old girl with myxoid liposarcoma. In children liposarcomas have a low-grade malignant potential.

On gross pathology most lipoblastomatous tumours were lobular, encapsulated and soft. Cut surfaces revealed colours of light yellow, tan, white, pink and red. Microscopically, fibrous connective septa divide the lipoblastoma into numerous lobules, with an eosinophilic mucinous to myxoid stroma. Mature adipocytes are located centrally and other adipocytes of varying stages of differentiation are noted randomly. The degree of differentiation is quite varied from primitive mesenchymal cells to spindled cells to lipoblasts to adipocytes [9] as seen in our lesion. Pleomorphism and cytological atypia are lacking, but mitotic activity may be seen. Hicks et al. [9] suggested that these lesions may differentiate to appear similar to a lipoma, which are typically composed of mature adipocytes, well circumscribed, surrounded by fibro-connective tissue capsule and lack lobulation.

Definitive treatment involves complete surgical excision of the tumour with clear margins. Local recurrence is likely if this unattainable, and whether the tumour is a circumscribed lipoblastoma or a diffuse infiltrative lipoblastomatosis. Recurrence rates reported by Chung and Ezinger were 14% [1], Collins and Chatten 9% [8] and 22% by Mentzel et al. [11]. In our case, this is most likely to happen, as the margins were not completely clear during surgical excision due to the diffusely infiltrative nature of the lesion.

Although lipoblastomatous tumours are benign tumours lacking the potential to metastasise, these neoplasms may cause a degree of morbidity due to the infiltrative nature of lipoblastamatosis on vital structures and visceral organs. Cottril et al. [13] recently reported a case of lipoblastomatosis arising in the heart and lung. Recognition of this tumour with proper classification into circumscribed lipoblastoma or diffusely infiltrative lipoblastomatosis is most important for surgical management and future review [9].

Acknowledgments

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