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| 原文題目(出處)： | Case report of buccal lipoblastomatosis in a 6-month-old infant. Int J Pediatr Otorhinolaryngol Extra 2009;4:6-9 |
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內文：

**Case report**

A 6-month-old Caucasian girl was referred for the assessment of **a small lump in the right cheek**. The mass has been increased since the age of 6 weeks, and it had begun to affect the infant's ability to breast-feed on the right side

Her medical history was unremarkable. She had not developed any primary dentition. Oral examination revealed a **2 cm lump in the right mucosa.**

- Non-ulcerated
- Incompressible
- Non-fluctuant
- No reported tenderness
- Facial nerve was deemed intact

**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 consistent with a **lipoblastoma**. The best management for this was **surgical enucleation**. 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.

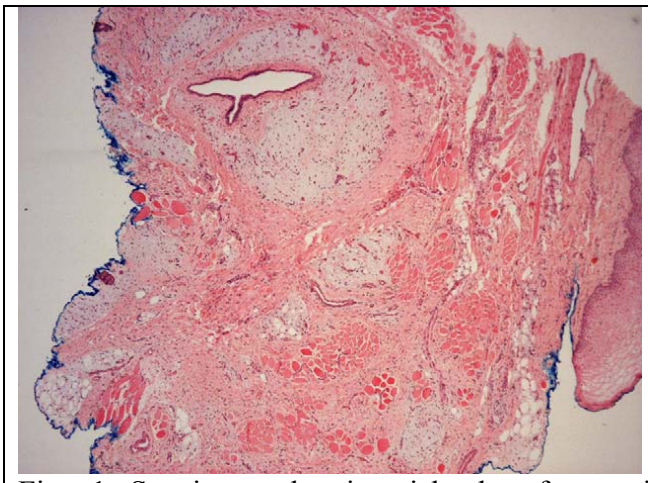

Fig. 1 Specimen showing islands of myxoid grey-white primitive adipose tissue extending through the stroma, with residual islands of skeletal muscle (40× magnification

Histopathological examination revealed a nodular lesion with a diffuse infiltrative margin extending into skeletal muscle

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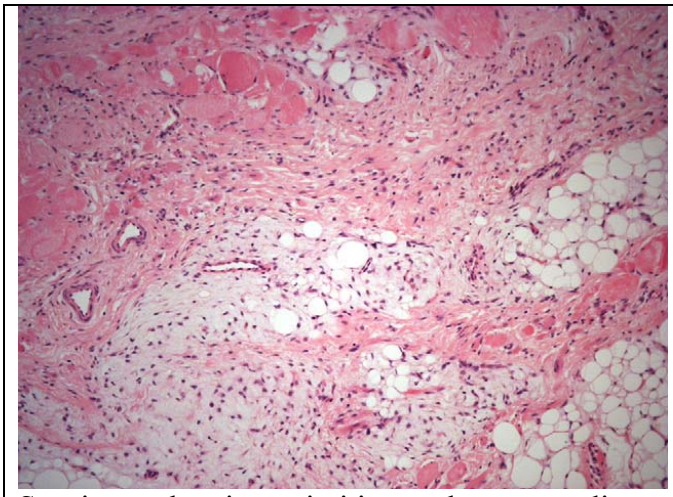
|  Specimen showing primitive and mature adipose tissue infiltrating into skeletal muscle (200× magnification). | The nodules were composed of lobules of mature and primitive adipose tissue with a myxoid appearance in places and delineated by fibrous septa. Numerous lipoblasts were present along with 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.

**Discussion**

Lipoblastomas are rare **benign tumours** arising from the fetal—**embryonal adipose tissue.**

- In **1926**, **Jaffe** 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** 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** 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.

**Current rate**

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

**Incident age**

These tumours predominantly occur in ***infants and young children under the age of 3 (90%)*** However, older patients have been recorded ageing from ***5 to 18 years old***

Incident site

- Lipoblastomas tend to occur in the superficial subcutis

- diffuse lipoblastomatosis is found deep in soft tissues, the mediastinum, retroperitoneum, thorax and abdomen

There have been **no reported cases of a lipoblastoma occurring in the buccal area**, although other rare sites have included the ***parotid gland***, ***eyelid*** and ***tonsillar fossa***.

- Case studies by Chung and Ezinger and Collins and Chatten ─ lipoblastomatous tumours are most likely to occur on the ***extremities*** as noted by ***72%*** of cases
- Collins and Chatten ─ **13 out of 18 cases** of lipoblastomatous tumours occurred on the **left** side of the body.

**Sexual predilection**

Lipoblastomatous tumours were more commonly found in **males than females** in many series, **(1.3~ 3.8 ratio)**, however in a conflicting report Hicks et al. stated there was **no gender predilection.**

**Clinical and histopathologic feature**

**Grossly**, 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
- ***eosinophilic*** mucinous to myxoid ***stroma***
- ***Mature adipocytes are located centrally and other adipocytes of varying stagesof differentiation are noted randomly.***
- Pleomorphism and cytological atypia are ***lacking***, but ***mitotic activity may be seen***.

**Diagnosis**

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

Differential diagnoses may include: sarcoma, neuroblastoma, germ cell tumour, lymphoma, epidermal inclusion cyst, lymphangioma, lipoma and Baker's cyst

**Treatment and recurrent rate**

Definitive treatment involves **complete surgical excision** of the tumor with clear margins. Local recurrence is likely if this unattainable, and whether the tumor is a circumscribed lipoblastoma or a diffuse infiltrative lipoblastomatosis. Recurrence rates reported by Chung and Ezinger were **14%**, Collins and Chatten **9%**, and **22%** by Mentzel et al.

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.**

**Conclusion**

Although these tumours are rare they should be included in differential diagnoses of soft tissue lesions in infants.

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| 1 | 下列何者的主要細胞來源是成熟的adipose tissue的過度增生<br>(A) lipoblastoma<br>(B) lipoma<br>(C) lipoblastomatosis<br>(D) lipoid proteinosis |
| 答案(B ) | 出處：Oral and Maxillofacial Pathology,Nevielle,Saunders W. B. Co.,1995,2nd edition |

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| 2 | 下列何者沒有local invasive的情形？<br>(A) ameloblastoma<br>(B) odontogenic myxoma<br>(C) lipoblastoma<br>(D) Pindberg tumor |
| 答案 (C) | 出處：Oral and Maxillofacial Pathology,Nevielle,Saunders W. B. Co.,1995,2nd edition |