The Journal of Laryngology & Otology (2014), ***128***, *212–215*.

doi:10.1017/S0022215113003630

CLINICAL RECORD

Infantile haemangiopericytoma: a rare congenital cervical tumour

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Abstract

Objective: Infantile haemangiopericytoma is a rare childhood vascular tumour of borderline malignancy and unpredictable clinical course. It can present a diagnostic challenge due to indeterminate clinical, radiological and pathological features. This report presents the case of a large congenital haemangiopericytoma of the neck in a neonate, and discusses diagnosis, imaging, pathology and surgical management.

Clinical presentation: A full-term neonate presented with a large posterior neck mass at birth. Pre-operative radiological appearances were suggestive of teratoma, but following surgical excision the diagnosis of infantile haemangiopericytoma was confirmed on histological analysis. There were no signs of recurrence at 12-month follow up.

Conclusion: Haemangiopericytoma can follow an aggressive course in adults, including local recurrence and metastasis. The infantile variant is rare but typically follows a distinct clinical course, and is associated with more benign behaviour compared with similar tumours in adults and children over one year. Congenital haemangiopericytoma can be effectively treated with surgery, without requiring adjuvant therapy.

Key words: Hemangiopericytoma; Congenital; Surgery; Neck

Introduction

Congenital neck masses represent a diverse group including branchial anomalies, lymphovascular malformations, and benign and malignant tumours. Many of these lesions, particularly branchial anomalies, do not manifest themselves until some time after birth, often years. Therefore, the differential diagnosis for a neonatal neck mass considers a specific subset of congenital cervical abnormalities the most common of which are vascular tumours and malformations, but which also includes dermoid cysts and teratomas. Neck masses in the neonate may be diagnosed prenatally on ultrasound and are a potential cause of neonatal airway problems. Vascular tumours and malformations are classified according to a system agreed by the International Society for the Study of Vascular Anomalies, which makes a clear distinction between tumours arising from vascular cellular hyperplasia, and malformations of capillary, venous, arteriovenous and lymphatic vessels resulting from aberrant morphogenesis.[1] Infantile haemangiopericytoma is a rare vascular tumour of borderline malignant potential. This report presents the diagnosis and management of a congenital infantile haemangiopericytoma of the neck in a neonate.

Case report

A female neonate born via normal vaginal delivery at full term (i.e. 40 weeks + 3 days) presented at birth with a large neck mass. This solid, pigmented mass measured 6 cm in maximum diameter and was located on the posterior neck, encroaching upon the border of the right trapezius muscle (Figure 1). Clinically, there was felt to be a vascular component to the mass. This mass had not been identified at antenatal screening. The child was otherwise well with no other abnormalities identified, a normal neurological examination, and no associated swallowing or airway problems. The child was transferred to our tertiary referral unit at 6 days of age for definitive management.

Computed tomography imaging identified an exophytic, soft tissue mass measuring $58 \times 43 \times 55$ mm (Figure 2). The mass was of soft tissue density but demonstrated some heterogeneity with an area of central, irregular calcification, suggestive of teratoma. There was moderate enhancement of the pseudocapsule but no significant central enhancement. There was no evidence of deep extension, and the plane between the mass and the paraspinal muscles was relatively well preserved.

Following multidisciplinary discussion with the oncology, paediatric otolaryngology, paediatric anaesthesia and paediatric radiology teams, the decision was made to proceed with surgical excision. The mass was completely excised 24 days post-natally without complications (Figure 3).

Following excision, the specimen was sent for histopathological examination. The tumour was found to be an encapsulated, well circumscribed mass. The densely

Accepted for publication 21 September 2013 First published online 10 February 2014

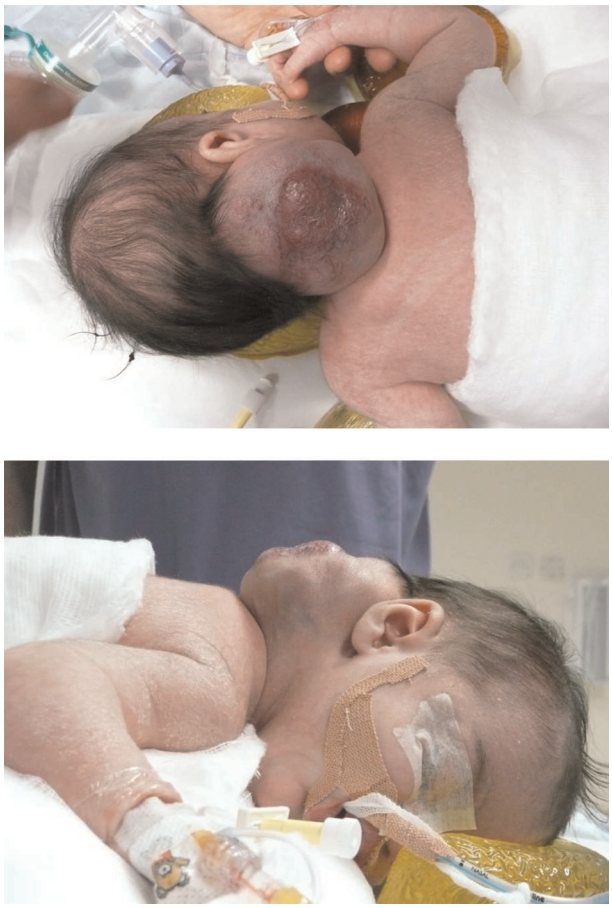

FIG. 1
Clinical photographs showing the pre-operative appearance of the neck mass in the 24-day-old neonate.

cellular lesion showed a multinodular growth pattern composed of interlacing sheets and whorls of spindle and round uniform cells. The cells were closely packed and

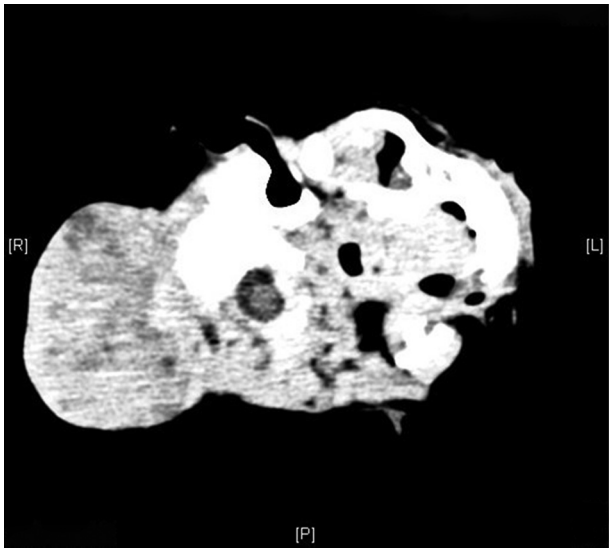

FIG. 2
Axial, contrast-enhanced computed tomography image demonstrating an exophytic, 58 × 43 × 55 mm, soft tissue mass extending from the right side of the neck. R = right; L = left; P = posterior

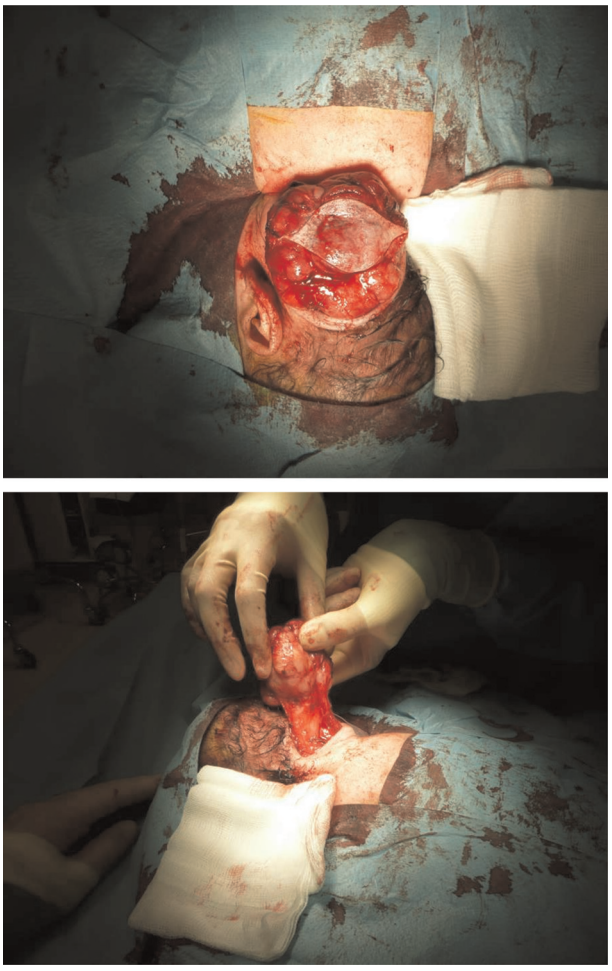

FIG. 3
Intra-operative photographs showing excision of the mass at 24 days of age.

showed medium-sized, vesicular nuclei with eosinophilic cytoplasm. The lesion was characterised by a prominent haemangioma pericytoma like vascular pattern showing

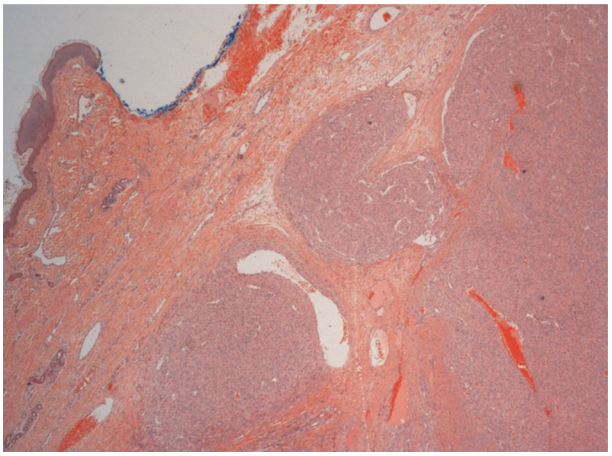

FIG. 4
Photomicrograph of the tumour demonstrating the typical multinodular arrangement of the tumour cells, with perivascular and intravascular satellitosis outside the main tumour lesion. (H&E; × 2.5)

https://doi.org/10.1017/S0022215113003630 Published online by Cambridge University Press

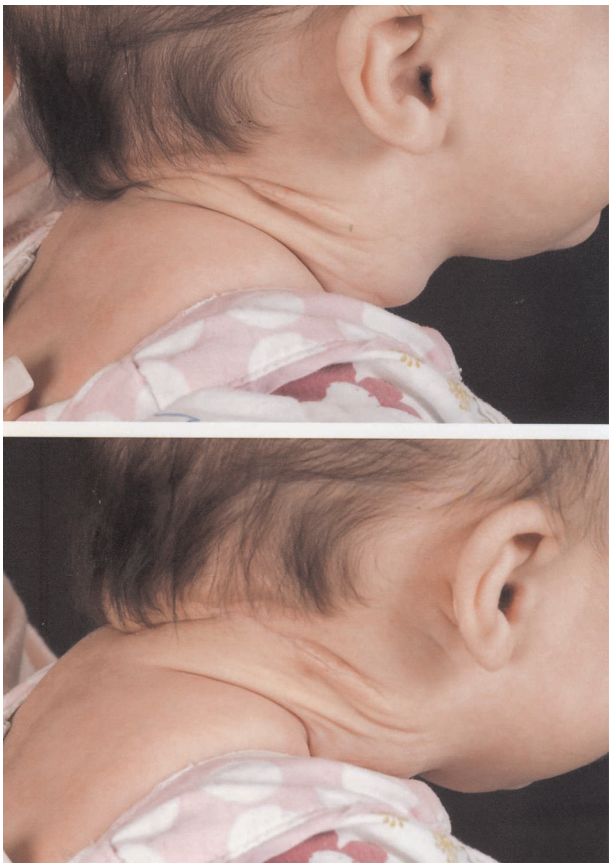

FIG. 5
Clinical photographs showing the post-operative appearance at two months.

numerous thick-walled, branching vessels with a staghorn configuration (Figure 4). No cellular atypia, pleomorphism or signs of malignancy were observed. The mitotic rate was low at 5/10 high-power fields (i.e. 5 mitoses in 10 different areas of the slide at high-power magnification). Genetic analysis and application of multiple tissue marker stains were performed, all of which were negative with the exception of vascular marker cluster of differentiation 34 protein, vimentin, integrase interactor 1, and factor VIII. Some areas of necrosis were noted. The differential diagnosis included benign vascular lesions and malignant soft tissue lesions including infantile fibrosarcoma and synovial sarcoma.

The case was discussed at our regional histopathology multidisciplinary team meeting; in the absence of a high mitotic rate, cellular atypia or other signs of malignancy, a final diagnosis of low grade infantile haemangiopericytoma was made. The lesion extended to the margins of the specimen but the exophytic nature of the specimen meant that the excision was considered to be complete.

The child recovered well and was discharged home 4 days post-operatively. Following review of the histology and discussion with the oncology multidisciplinary team, it was decided that no further treatment was required. The child continued to be monitored in the out-patient clinic (Figure 5). At 12 months, there were no signs of recurrence. At the time of writing, following advice from the regional oncology service, we planned to keep the child under surveillance for five years.

Discussion

Haemangiopericytoma is a rare vascular tumour first described by Stout and Murray in 1942.[2] Originally thought to arise from the pericytes that surround vessels, the exact nature of the lesion remains controversial, and it is now considered by many to be part of a spectrum of myopericytoma lesions related to infantile myofibromatosis.[3] It can follow an aggressive course, including local recurrence and metastasis to bone, lung and liver. Presentation in children is rare, accounting for only 5–10 per cent of cases.

Infantile haemangiopericytoma is a rare variant which may present at birth or in the first year of life, and which tends to occur in the head and neck.[4] The infantile variant typically follows a distinct clinical course, with a more benign behaviour than in adults or children over one year,[5] and is considered to be a distinct entity. A conservative surgical approach is therefore recommended in infants.[6]

Congenital tumours may be identified on antenatal ultrasound.[4,7] Pre-operative diagnosis presents a challenge as clinical and radiological features may be indistinct and insufficient to distinguish the lesion from other soft tissue tumours.[8] The differential diagnosis includes other soft tissue tumours of benign, malignant or indeterminate biological behaviour, including angiosarcoma and neuroblastoma. It is difficult to distinguish these lesions clinically, and diagnosis relies on histological findings. Given the vascular nature of the tumour and the associated risk of bleeding, complete excision may be preferable to less invasive biopsy methods. Ultrasound and Doppler investigation may be used to assess vascularity prior to biopsy.[9]

In the presented case, histological diagnosis was aided by the typical appearance of spindle cell fascicles with eosinophilic cytoplasm and a pericytoma-like vascular pattern in staghorn configuration. Haemangiopericytoma is not thought to be associated with any particular immunohistochemical staining profile, though typically lesions are positive for the vascular marker cluster of differentiation 34 protein, and also for vimentin and cluster of differentiation 99 protein.[10,11] The features of standard proliferative markers do not fully correlate with the clinical course, though histological features such as necrosis, mitoses, vascular invasion and cellular pleomorphism are associated with more aggressive tumour behaviour in adults.[12,13] However, in infants features such as mitotic figures and focal necrosis are not necessarily predictive of malignant tumour behaviour,[6] and the course of infantile haemangiopericytoma is generally benign. In the presented case, the differential diagnosis of infantile fibrosarcoma was considered; however, molecular genetic studies did not identify any translocation, and the vascular pattern and the strong positivity for cluster of differentiation 34 protein were suggestive of the final diagnosis.

- **Haemangiopericytoma is a rare tumour of indeterminate nature**
- **Presentations in infancy tend to be benign, but later presentations can be malignant**
- **A case of neonatal congenital haemangiopericytoma in the neck is presented, with successful surgical management**

Where resectable, surgical excision is associated with a good prognosis and a low recurrence rate. Where surgical excision

https://doi.org/10.1017/S0022215113003630 Published online by Cambridge University Press

is not possible, neoadjuvant chemotherapy has been used successfully[14] and has been associated with maturation to haemangioma.[5]

Conclusion

This case highlights the challenge presented to clinicians by these rare congenital vascular tumours: clinical and radiological features may be unable to establish a clear diagnosis. Initial management decisions, including the decision to surgically excise the tumour, may need to be taken before the histological nature of the lesion has been confirmed. The histopathological features may suggest malignancy, leading to over-treatment of these tumours, although a benign course is expected in infants.

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Ms E M Farrar takes responsibility for the integrity of the content of the paper
Competing interests: None declared