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Giant congenital exophytic strawberry-like mass in a newborn

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Clinical findings

A male newborn was referred at birth because of a suspected giant congenital haemangioma on his back. He had been delivered vaginally at 38 + 5 weeks after a normal pregnancy. Ultrasonography during the fifth month of pregnancy had not detected the mass; however, at birth, a purplish-red, reniform, nodular and pedunculated skin tumour measuring 80 × 50 × 35 mm was visible on the right paravertebral lumbosacral area. This strawberry-like mass had a tense consistency and a smooth surface with homogeneous, diffuse and yellowish point depressions (Fig. 1a,b). Doppler ultrasonography showed that the tumour was not vascular in nature. Complete surgical excision was performed and the tumour was sent for histology.

Histopathological findings

Histological examination showed a well-circumscribed dermal lesion composed of multiple folliculosebaceous units, with frequent cystic dilated hair infundibules (Fig. 2a). The units were embedded in a loose fibromyxoid stroma with interspersed thin smooth muscle bundles, which were nicely highlighted by desmin. Small lobules of immature adipose tissue were scattered within the stroma (Fig. 2b). Small nerve trunks and ectatic vessels were also seen (Fig. 2c). No connections between the sebaceous lobules and the overlying epidermis were noted. At the periphery, a stromal cleft separated the lesion from the surrounding dermis.

What is your diagnosis?

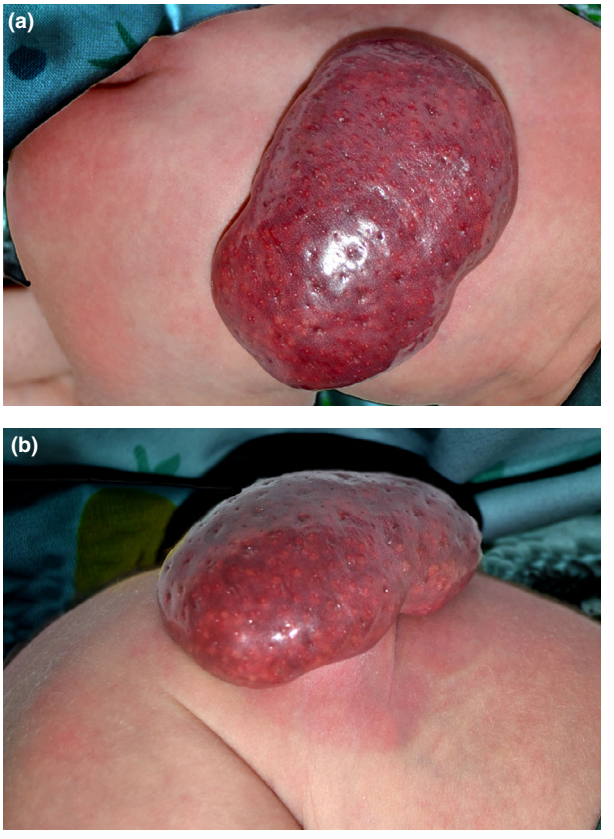

Figure 1 (a,b) Strawberry-like, reniform and pedunculated skin tumour measuring 80 × 50 × 35 mm on the right paravertebral lumbosacral area.

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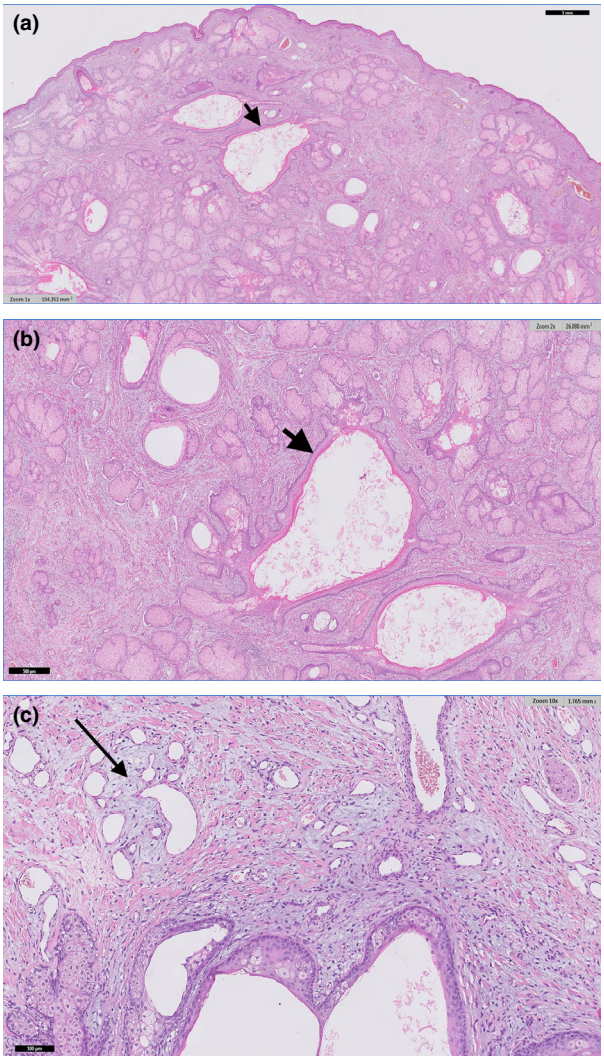

Figure 2 (a,b) Dermal proliferation of sebaceous glands with some cystic folliculosebaceous units (arrowhead); (c) thin bundles of muscle fibres, small lobules of immature adipose tissue (arrow), small nerves and vessels were visible. Haematoxylin and eosin, original magnification (a) × 10; (b) × 20; (c) × 100.

Diagnosis

Folliculosebaceous cystic hamartoma (FSCH).

Discussion

FSCH, first described in 1991 by Kimura *et al.*,[1] is an uncommon benign skin lesion, with < 100 cases reported in the literature since the first description. FSCH usually manifests with a single, small (5–15 mm), flesh-coloured, sessile or pedunculated mass, frequently located on the head, in particular on the central part of the face and nose. FSCH is usually isolated, and a syndromic form has never been reported in the literature. It primarily affects middle-aged adults,[1–2] but has also been described in infancy and a very few congenital cases have been reported.[3–5] Emsen and Livaoglu described a case of FSCH appearing at 6 months of age as a congenital case.[6]

According to previous reports, FSCH is defined 'giant' when the lesion ranges from 50 to 230 mm in diameter,[2] and only a very small number of giant FSCH have been reported.[2–6] Congenital FSCH were described as 'giant' in only three cases, with the patients being 6, 9 and 12 months of age; however, the lesions were smaller at birth in all three cases.[3–5] To our knowledge, our patient is the first case of congenital FSCH that was giant since birth.

FSCH is considered a hamartoma because of the coexistence of multiple mature ectodermal and mesodermal elements. Histological examination is necessary to define the diagnosis. Whereas the clinical aspects and the localization are characteristic in adults, there are variations in its appearance in newborns and children. CO_2 laser, oral acitretin and surgery are the most useful therapeutic options.[1–6] Our patient was treated with excision surgery, and has been in good health since then.

Acknowledgement

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Learning points

- FSCH is an uncommon hamartoma.
- It is usually seen in middle-aged adults and rarely described in children.
- Congenital FSCH has been reported in very few cases, and giant congenital FSCH is an extremely rare entity.
- Histopathology is necessary to establish the diagnosis.

References

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CPD questions

Learning objective

To gain up-to-date knowledge on the classification of and investigations for folliculosebaceous cystic hamartoma.

Question 1

Why is folliculosebaceous cystic hamartoma (FSCH) defined as a hamartoma?

(a) Because of the coexistence of multiple mature ectodermal and mesodermal elements.
(b) Because of the strawberry-like appearance.
(c) Because FSCH is a congenital tumour.
(d) Because FSCH is a benign tumour.
(e) Because of the proliferation of multiple sebaceous glands.

Question 2

What is the most useful investigation for diagnosis of folliculosebaceous cystic hamartoma (FSCH)?

(a) Clinical examination.
(b) Magnetic resonance imaging.
(c) Ecocolour Doppler.
(d) Histology.
(e) Dermoscopy.

Instructions for answering questions

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- Reflect on the article.
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