A Subcutaneous Tumour on the Posterior Neck: A Quiz

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A 51-year-old healthy Japanese man presented with a 1-year history of a subcutaneous asymptomatic tumour, 6×5 cm in size and gradually increasing, on the posterior neck. The patient had undergone cupping therapy on the shoulder and neck for alleviating stiffness once a week for 20 years. On physical examination an elastic, hard, immobile mass with no tenderness or redness was palpable on the posterior neck (Fig. 1A) and a rounded purpura was observed on the shoulder, possibly caused by the cupping therapy. Laboratory test results including blood count, biochemistry and coagulation were within normal limits. Magnetic resonance imaging (MRI) showed a well-circumscribed, rounded solid mass, with homogeneous intermediate signal intensity (isointense to muscle) on T1-weighted images (T1-WI) (Fig. 1B) and inhomogeneous mild high signal intensity involving sporadic scattered low signal intensity areas inside on T2-weighted images (T2-WI) (Fig. 1C) and short T1 inversion recovery (STIR) (Fig. 1D).

What is your diagnosis? See next page for answer.

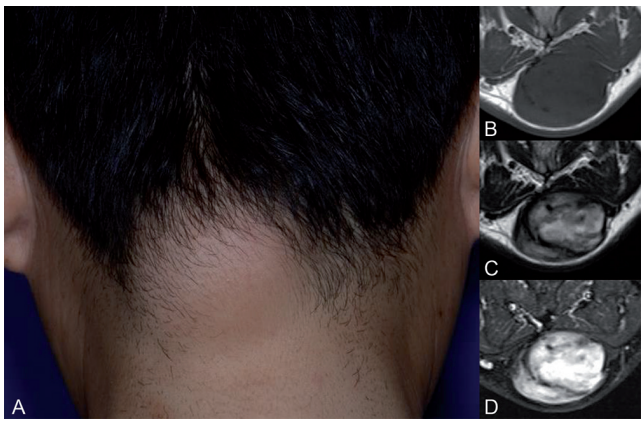

Fig. 1. (A) A 6×5 cm asymptomatic subcutaneous tumour on the posterior neck. (B, C, D) Magnetic resonance imaging (MRI) revealed homogenous, intermediate signal intensity on (B) T1-weighted image, and inhomogeneous high signal intensity with scattered low signal intensity areas on (C) T2-weighted and (D) short T1 inversion recovery (STIR).

doi: 10.2340/00015555-2972
Acta Derm Venereol 2018; 98: 817–819

ANSWERS TO QUIZ

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Diagnosis: **Desmoplastic fibroblastoma (DFB)**

Based on MRI findings and the history of cupping therapy on the shoulder and neck, an expanding haematoma was initially suspected. The course of the tumour without treatment was observed carefully, with the expectation of spontaneous resolution within several months. However, no tendency to resolution was seen, and thus the tumour was totally resected. Histological analysis revealed an oval-shaped, well-circumscribed, subcutaneous tumour (Fig. 2A), consisting of bland fibroblastic cells without atypia, sparsely distributed, with a rich stromal component (Fig. 2B, C). These fibroblastic cells stained positively for vimentin (Fig. 2D) and negatively for S100 protein, epithelial membrane antigen, CD34, α-smooth muscle actin, desmin, cytokeratin AE1/AE3, and mucin 4, which suggested a diagnosis of desmoplastic fibroblastoma (DFB). To confirm this diagnosis, FOS-like antigen-1 (FOSL1) immunohistochemical staining was performed, as reported previously (1). This revealed a diffuse, strong FOSL1 nuclear immunoreactivity (Fig. 2E) compared with the surrounding area outside the tumour (Fig. 2F). No recurrence was found during 1-year follow-up.

DFB is a benign fibroblastic/myofibroblastic soft tissue tumour which may occur in various locations of the body (2–5). DFB is generally located in the deep subcutaneous tissue, fascia or skeletal muscle, and occurs predominantly in males (4:1), with a median age of presentation of 50 years (5). The reported diameters of DFB are 1–20 cm, with a mean of 3 cm (5). The recommended treatment is complete surgical excision (5, 6).

Histologically, DFB is composed of fibroblasts/myofibroblasts immersed in an abundant dense collagen with variable myxoid stroma, and histological differential diagnosis includes a wide variety of soft tissue tumours (7). A definite diagnosis can be made in combination with immunohistochemical staining, although diagnosis is sometimes difficult. Recently, microarray analysis revealed increased mRNA expression of FOSL1 in DFBs due to 11q12 rearrangement (7), and DFB specifically shows strong FOSL1 nuclear immunoreactivity (1). FOSL1 is therefore a useful tool for differentiating DFB from other histological mimics.

DFB has a varying intensity on MRI; intermediate intensity to low signal intensity on T1-WI and low to high signal intensity on T2-WI (6, 8–10). Interestingly, low signal intensity areas on T2-WI correspond to abundant collagen fibres and low cellularity, while high signal intensity areas on T2-WI represent dense fibroblast cells, loose collagen fibres, and abundant myxoid areas (8). In our case, the centre of the tumour consisted of dense fibroblast cells, loose collagen fibres, and abundant myxoid areas, which could reflect a high signal intensity area on STIR and T2-WI (Fig. 3A). On the other hand, the peripheral area of the tumour consisted of hypocellular component with dense collagen fibres corresponding with low signal intensity on both STIR and T2-WI (Fig. 3B). Thus, DFB has a variable cellularity and collagen fibre density, depending on each case, and therefore, as in the current case, it is often difficult to distinguish DFB from other subcutaneous tumours by MRI alone.

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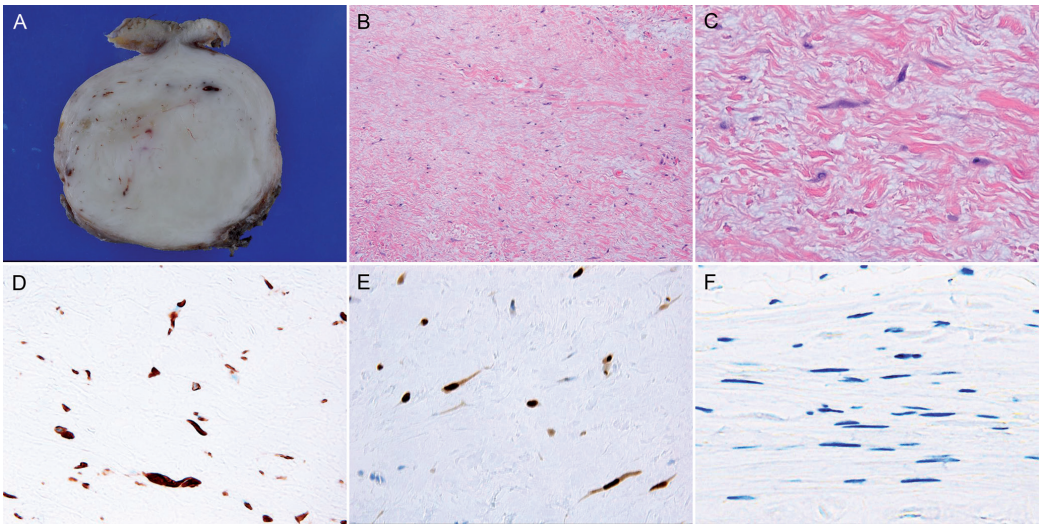

Fig. 2. (A) Macroscopic appearance of a resected, well-circumscribed, subcutaneous tumour. (B, C) Representative images of haematoxylin-eosin staining of the subcutaneous tumour (B, ×100; C, ×400). (D) Immunohistochemical staining for vimentin in the tumour. (E, F) Immunohistochemical staining for FOS-like antigen-1 in the tumour and the area surrounding the tumour. Strong positivity for FOS-like antigen-1 was observed in the nuclei of spindle cells in the tumour (E, ×400) compared with the surrounding area (F, ×400).

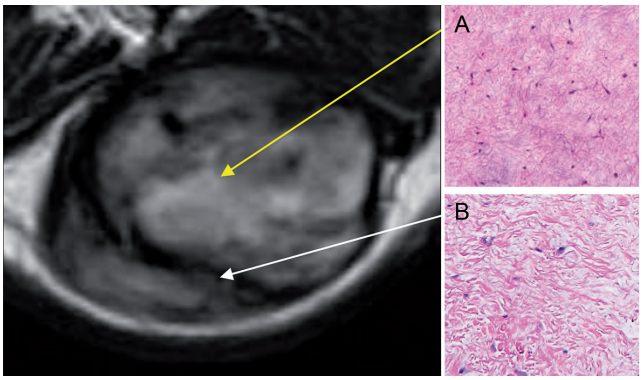

Fig. 3. (A) The centre of the tumour consisted of relatively dense fibroblast cells, loose collagen fibres, and abundant myxoid areas, which could correspond with a high signal intensity area on T2-weighted image (*yellow arrow*). (B) The peripheral area inside the tumour consisted of hypocellular component with dense collagen fibres corresponding with a low signal intensity area on T2-weighted image (*white arrow*).

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