Spitz naevus was first described by Sophie Spitz in 1948 (1). It is a variant of benign melanocytic naevus composed of spindled and epithelioid melanocytes, found predominantly in children and adolescents. A number of histopathological variants have been reported, such as desmoplastic, hyalinized, angiomatous, granulomatous, pagetoid, myxoid, tubular and rosette forms (2). We report here a case of tubular Spitz naevus with variable histopathological images, which partly resembled ependymal rosette in neurogenic neoplasms and partly resembled eccrine spiradenoma.

CASE REPORT

A 9-year-old boy presented with an asymptomatic red dish nodule, which had been present for 2 months. On clinical examination, a pink-red, elastic hard nodule with papillomatous surface, 8 mm in diameter, was noted on his left forearm (Fig. 1). He was otherwise healthy, and no other specific skin eruptions were found. Pseudolymphoma, xanthogranuloma and Spitz naevus were considered clinically. Since the eruption had not changed over a 3-month period careful observation, we excised it for histopathological diagnosis. In the dermo-epidermal junction, spindle cells with faint melanins were aggregated to form nests (Fig. 2a). Small nests of tumour cells showed some clefts (Fig. 2b). Giant cells and dense lymphocytic infiltration were also observed. In the entire dermis, round-to-oval epithelioid cells with eosinophilic cytoplasm were remarkably proliferated. Interestingly, the majority of such tumour cells formed microtubular appearance, variably mixed with microvénules (Fig. 2c). A mass of basophilic tumour cells with microcystic appearance was also observed in the subcutis, mimicking eccrine spiradenoma (Fig. 2d). Regardless of the diverse appearance of tumour cells, these cells had a uniform immunohistochemical profile; S-100+, Melan-A+, HMB-45– and AE3/AE1– (Fig. 2e and f). From these clinico-pathological findings we diagnosed the patient with tubular Spitz naevus. After excision no local recurrence or metastasis has been observed to date.

DISCUSSION

Tubular Spitz naevus, a very rare variant, was first described by Burg and colleagues in 1998 (3). They reported a case with a characteristic finding, the formation of tubular structures bordered by S-100+ epithelioid cell strands. Confocal laser scan microscopic analysis revealed tubular and/or microcystic spaces formed by tumour cells, at least 30 μm long. Requena et al. (2) investigated 349 lesions of Spitz naevi, and detected only one tubular type case.

The pathomechanisms and clinical significance of tubular Spitz naevus are unknown. Burg et al. (3) speculated that the tubular images might be caused by apoptosis of centrally located naevus cells or secretion of autocrine or paracrine factors. Conversely, Ziemer et al. (4) reviewed 31 Spitz naevi and found “tubular” structures in 15 cases, and they suggested that such tubular appearance may result from retraction of aggregated epithelioid tumour cells, secondary to fixation of formalin. As a similar histopathological variant, Kantrow and colleagues (5) reported a case of Spitz naevus with rosette-like formation. They pointed out that the rosettes resembled Homer-Wright rosettes in neurogenic tumours, such as neuroblastoma, characterized by a radial arrangement of cells with centrally situated fibrillar material composed of neuropil. In our case, unlike Homer-Wright rosettes, tumour structures contained apparent empty-appearing lumen and lacked fibre-rich neuropil or central cytoplasmic projections. Such tubular patterns partly resembled “true ependymal rosette” observed in well-differentiated ependymoma (6). Kantrow et al. (5) speculated that shared neural crest origin might induce similar characteristic images to neurogenic tumours, which also seemed to explain our case. Indeed, rosette-like, tubular or glandular patterns are observed in other melanocytic neoplasms, such as...
Clark’s naevus and malignant melanoma (7, 8). Taking these observations into consideration, tubular, rosette and glandular structures seen in Spitz naevus might reflect declined differentiation to neurological lineages from neural crest, and the degree of artefacts caused by formalin fixation can explain histopathological differences in these structures.

Since dense basophilic tubular structures were observed in the subcutis mimicking eccrine spiradenoma, we first considered the case might be Spitz naevus with sweat duct differentiation or concomitant sweat duct tumours, such as spiradenoma and eccrine angiomatous hamartoma. Recently, a case of Spitz naevus in concordance with a syringoma was reported, which might support a possible co-occurrence of Spitz naevus and eccrine duct neoplasms (9). However, in our case, the positivity of S-100 and negativity of AE1/AE3 in the tubular areas denied the concomitant sweat gland neoplasms. Although the significance of the rare histological subtype is unknown, dermatologists and pathologists should avoid potential misdiagnosis caused by the wide variety of histopathological images in Spitz naevus.

The authors declare no conflicts of interest.

REFERENCES