2. Finck FM, Schwinn CP, Keasbey LE. Clear cell hidradenoma of the breast. *Cancer* 1968; 22: 125–35.
3. Ohi Y, Umekita Y, Rai Y, *et al*. Clear cell hidradenoma of the breast: a case report with review of the literature. *Breast Cancer* 2007; 14: 307–11.
4. Gorunova L, Mertens F, Mandahl N, *et al*. Cytogenetic heterogeneity in a clear cell hidradenoma of the skin. *Cancer Genet Cytogenet* 1994; 77: 26–32.
5. Behboudi A, Winnes M, Gorunova L, *et al*. Clear cell hidradenoma of the skin—a third tumor type with a t(11;19)-associated TORC1-MAML2 gene fusion. *Genes Chromosomes Cancer* 2005; 43: 202–5.
6. Winnes M, Mölne L, Suurküla M, *et al*. Frequent fusion of the CRTC1 and MAML2 genes in clear cell variants of cutaneous hidradenomas. *Genes Chromosomes Cancer* 2007; 46: 559–63.
7. Kazakov DV, Vanecek T, Belousova IE, Mukensnabl P, Kollertova D, Michal M. Skin-type hidradenoma of the breast parenchyma with t(11;19) translocation: hidradenoma of the breast. *Am J Dermatopathol* 2007; 29: 457–61.
8. Domoto H, Terahata S, Sato K, Tamai S. Nodular hidradenoma of the breast: report of two cases with literature review. *Pathol Int* 1998; 48: 907–11.
9. Mote DG, Ramamurti T, Naveen Babu B. Nodular hidradenoma of the breast: A case report with literature review. *Indian J Surg* 2009; 71: 43–5.
10. Vasconcelos I, Radke C, Tellechea Ó, Schoenegg W. Hidradenoma of the breast: a source of diagnostic confusion. *Breast J* 2015; 21: 681–2.
11. Sehgal S, Goyal P, Ghosh S, Mittal D, Kumar A, Singh S. Clear cell hidradenoma of breast mimicking atypical breast lesion: a diagnostic pitfall in breast cytology. *Rare Tumors* 2014; 6: 5343.
12. McNiff J, Vassallo C, McCalmont TH, *et al*. Benign tumours with apocrine and eccrine differentiation. In: LeBoit PE, Burg G, Weedon D, Sarasin A, editors. *WHO Classification Skin Tumours*. Lyon: WHO, 2006; 139–48.
13. Miettinen M, McCue PA, Sarlomo-Rikala M, *et al*. Sox10–a marker for not only schwannian and melanocytic neoplasms but also myoepithelial cell tumors of soft tissue: a systematic analysis of 5134 tumors. *Am J Surg Pathol* 2015; 39: 826–35.
14. Hsieh MS, Lee YH, Chang YL. SOX10-positive salivary gland tumors: a growing list, including mammary analogue secretory carcinoma of the salivary gland, sialoblastoma, low-grade salivary duct carcinoma, basal cell adenoma/adenocarcinoma, and a subgroup of mucoepidermoid carcinoma. *Hum Pathol* 2016; 56: 134–42.

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## Melanotic neuroectodermal tumour of infancy presenting as an undifferentiated round cell tumour in the soft tissue of the forearm

Sir,

Melanotic neuroectodermal tumour of infancy (MNTI), first described by Krompecher in 1918 as ‘congenital melanocarcinoma’, is a rare tumour arising mainly in infants under 1 year of age. Various names such as pigmented ameloblastoma, melanotic adamantinoma, melanotic progonoma, melanotic epithelial odontoma, pigmented teratoma, pigmented congenital epulis, and retinal anlage tumour have been used to describe this tumour, illustrating the uncertainty of its origin and histogenesis.[1] Based on modern ultrastructural, immunohistochemical and electron microscopic studies, MNTI is now widely accepted to be of neural crest origin.[2] The currently used term MNTI was introduced by Borello and Gorlin.[3] MNTI has a preferred occurrence in the head and neck region, with the maxilla, mandible and skull as the main sites of origin.[4] Occasionally, other sites, such as the epididymis, mediastinum, brain and skin may be involved, and there are rare reported cases of MNTI arising in the soft tissue of the extremities.[5] Although MNTI is characterised by a relatively benign clinical course and is usually cured by a wide local excision, its rapid onset and alarming local growth are often clinically worrisome. Moreover, MNTI has the capacity for local destruction and studies have reported recurrence and metastatic spread in 10–15% and 3% of cases, respectively.[6] The potential for recurrence or metastasis cannot be predicted by the histopathological characteristics of the tumour. In this report, we present the case of a 3-month-old male infant with an MNTI located at the forearm, successfully treated with neoadjuvant chemotherapy and surgical resection. We discuss the diagnostic difficulties caused by its very uncommon location and its unusual histological appearance. Furthermore, this case also demonstrates the effects of chemotherapeutic treatment on the histopathological appearance of this paediatric tumour.

A 3-month-old boy was referred to our hospital in January 2013 for evaluation of a rapidly growing tumour in the left forearm, which had been noticed by his parents 2 weeks previously. There were no abnormalities reported at birth. The infant was otherwise healthy, and his growth and development were adequate for his age. On palpation, a firm and mildly tender subcutaneous nodule of about 3 cm was noticeable in the left forearm. The overlying skin was intact and freely movable, but revealed prominent telangiectasia. An incisional biopsy was performed. Microscopy showed a poorly differentiated neoplasm composed of clusters of monomorphic small rounded cells with relatively uniform rounded nuclei and scant cytoplasm. The tumour cells were surrounded by a prominent desmoplastic stroma (Fig. 1A–C). Mitotic figures were infrequently seen and necrosis was absent. Immunohistochemistry showed positivity for pan-cytokeratin AE1/AE3 and CAM 5.2 in scattered tumour cells, while staining for NSE, EMA, desmin, myogenin (MYF4), CD99, CD45, S100, WT1, ERG, CD34, synaptophysin, chromogranin, HMB45 and Melan A were negative (Fig. 1D). SMARCB1 (INI-1) expression was preserved in the tumour cell population. Given the growth pattern and positivity for keratins, we wanted to exclude an unusual example of a desmoplastic small round cell tumour (DSRCT) arising in a limb. Fluorescence *in situ* hybridisation (FISH) analysis showed no presence of *EWSR1* gene rearrangement.

Based on the uncommon histopathological findings in the biopsy, the tentative diagnosis of an unclassified round cell malignant neoplasm was made. For treatment purposes, the tumour was regarded as an undifferentiated sarcoma. Because of the size and location of the tumour, an immediate surgical treatment was contraindicated. Instead, the patient was treated according to the paediatric non-metastatic rhabdomyosarcoma (RMS) 2005 protocol (standard risk group, subgroup D), with a chemotherapy regimen based on ifosfamide (I), vincristin (V) and actinomycin (A). As soon as the patient reached the age of 6 months, doxorubicin was added to the regimen. The chemotherapy resulted in a significant reduction of the tumour size. Seventeen weeks after the initial IVA administration, a wide excision was performed with successful removal of the tumour.

Macroscopically, the tumour was well demarcated but not encapsulated. The cut surface exhibited a striking black appearance and the texture was tough (Fig. 2A). Microscopic examination of the surgical specimen showed a totally different histological appearance compared to the pretreatment biopsy sections. The tumour was composed of large epithelioid to polygonal cells arranged in trabeculae, tubules and pseudoglandular (pseudoalveolar) structures, embedded

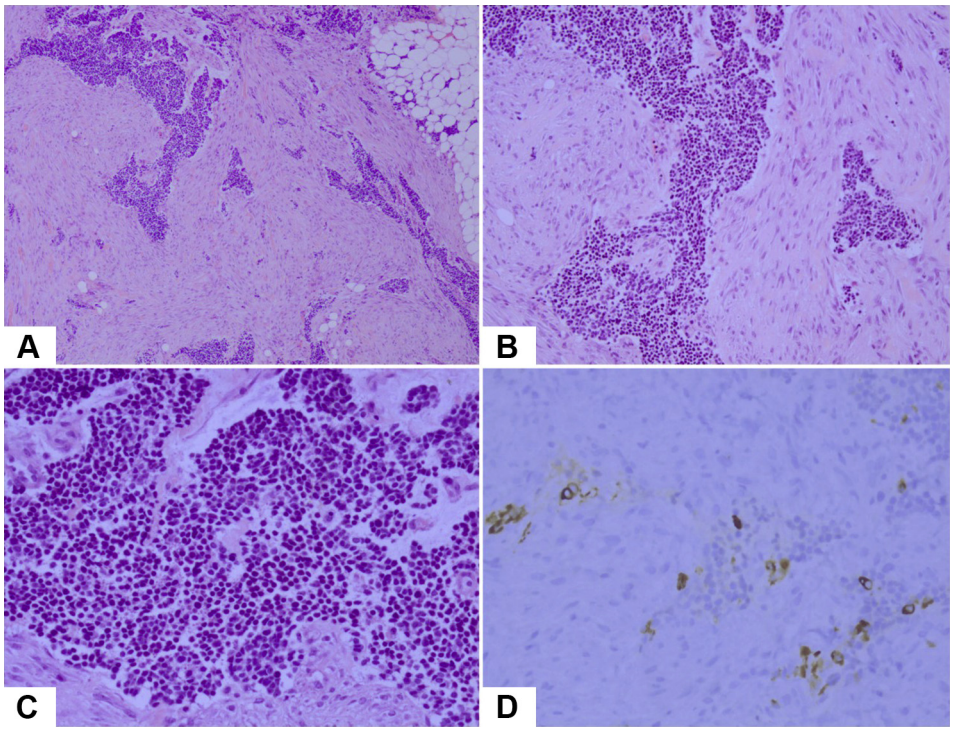


**Fig. 1** Incisional biopsy. (A,B) Microscopy showed a poorly differentiated neoplasm composed of clusters of monomorphic small rounded cells, surrounded by a prominent desmoplastic stroma (H&E). (C) Monomorphic small rounded cells with relatively rounded nuclei and scant cytoplasm. (D) Immunohistochemistry showed positivity for pancytokeratin AE1/AE3 in scattered tumour cells.

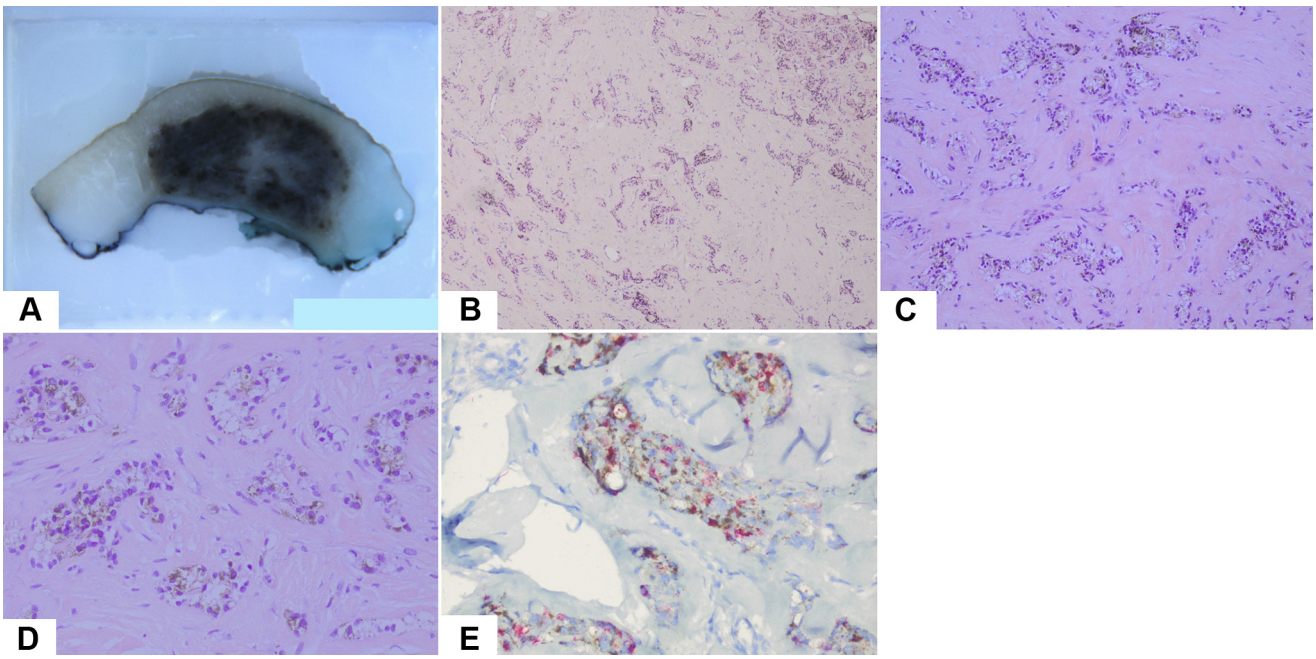


**Fig. 2** Surgical excision specimen. (A) Serial sections showed a well-circumscribed, not encapsulated tumour that exhibited a striking black appearance. (B,C) Microscopically, the tumour was composed of large epithelioid to polygonal cells arranged in trabeculae, tubules and pseudoglandular structures, embedded in a prominent, well-vascularised desmoplastic stroma (H&E). (D) The large epithelioid cells had abundant eosinophilic cytoplasm and contained visible particles of black melanin pigment (H&E). (E) The neoplastic cells were strongly positive for HMB45.

in a prominent, well-vascularised desmoplastic stroma (Fig. 2B,C). These large epithelioid cells had abundant eosinophilic cytoplasm and contained visible particles of black pigment (Fig. 2D). The pigment stained with the histochemical Masson Fontana stain and was identified as melanin. Immunohistochemically, the neoplastic cells were strongly positive for pancytokeratin AE1/AE3, synaptophysin, HMB45 and NSE (Fig. 2E). Interestingly, the undifferentiated small blue round cell component, which was the predominant component in the pretreatment biopsy, was not observed in this surgical excision. Based on histomorphology and immunoprofile, the tumour was diagnosed as MNTI of the left forearm. All the section margins were free of tumour. The original biopsy was revised. Although one