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Subcutaneous Sacrococcygeal Myxopapillary Ependymoma. A Case Report

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SUBCUTANEOUS SACROCOCCYGEAL myxopapillary ependymomas are very rare neoplasms, occurring as primary tumours of the skin and soft tissues of the sacrococcygeal area, without demonstrable connection with the spinal cord [1]. In this case, it is believed that the coccygeal medullary vestige is the source of ependymal cells, which later develop into the neoplasm [1]. After the original description by Mallory in 1902 [2], only about 40 cases have been described in the literature [1, 3–5], affecting both sexes equally [1, 3, 4], with a mean diagnostic age of 17 years [1, 3]; the pilonidal area is the most frequent site of the tumour, with buttock the second [1, 3, 5]. We present here the case of a child (10 years, 5 months old) with a subcutaneous sacrococcygeal myxopapillary ependymoma, localised both in the pilonidal area and left buttock. This is the first case report with these characteristics in the literature. The patient was referred to the "C" Surgery Department at the National Cancer Institute of Naples, because of a nodular lump in the soft tissues of the intergluteal fold, overlying the sacrum and the coccyx. This lump, noted at birth, had increased sharply in the preceding 4 months, reaching about 7 cm at its largest diameter. Clinical and instrumental diagnosis of "lipoma" was made, and wide and easy excision was performed. At surgery, a tumour was found in the soft tissues of the sacrococcygeal area and left buttock, without involvement of the coccygeal fascia and underlying muscle. On microscopic examination of paraffin sections, the tumour entirely revealed a papillary architecture. The papillae had a myxopapillary configuration, typical of cauda equina ependymomas. The myxoid material in the bulbous papillae appeared as a very fine red network at the ependymal cell wall when stained by the periodic acid–Shiff (PAS) techique. To identify both glial fibrillary acid protein (GFAP) and S-100 antigen, an immunohistochemical method (peroxidase anti-peroxidase technique) was performed on paraffin sections of the tumour. With GFAP, the ependymal tumour cells stained positively, although the reaction was not always uniform (Figure 1). The immunoreactivity for S-100 antigen was strong within the normal small nerves in the stroma, but weak or negative within the ependymal tumour cells. These immunohistochemical findings are in agreement with the literature [1]. To date, no correlation between clinical presentation of the mass and prognosis has been described. Unfortunately, we suspect that large tumour size (about 10 cm at the largest diameter) and the contemporaneous tumour localisation in two different sites (pilonidal area and buttock) may represent poor prognosis factors. However, the young age of our patient must be considered: it was seen, in fact, that rare cases of metastases have all

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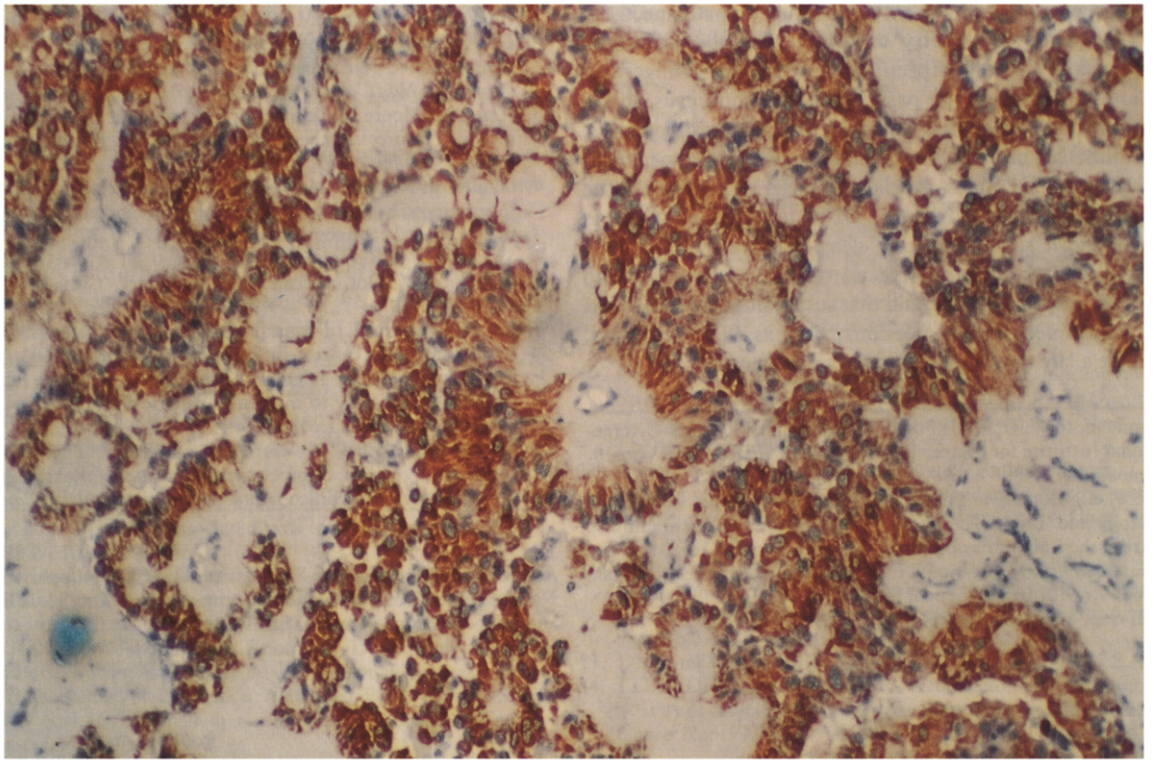

Fig. 1. Diffuse and strong staining of numerous ependymal tumour cells with anti-GFAP (×100 magnification).

been documented in adult patients [1, 6].

All previous authors agree that long-term follow-up is necessary [1, 5–7]. Metastases, in fact, occur in 20% of cases, 5 to 15 years after diagnosis [1], and their localisation in the lung after a 20-year follow-up has been described in one case [6]. However, no correlation between tumour cytohistological patterns (papillary, solid, myxoid) and clinical development has been documented [1, 5, 7]. In only one case with pulmonary metastases were tumour cells identified in single blood vessels of the primary specimen [1]. More frequent sites of metastases are lung and inguinal lymph nodes [1, 6]. For these reasons, yearly clinical examinations and chest X-ray should be performed, even if, as in the case presented here, no tumour cells are evident in the blood vessels.

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Correction—Evidence for abrogation of oncogene-induced radioresistance of mammary cancer cells by hexadecylphosphocholine *in vitro*. This paper by E.A. Bruyneel *et al.* appeared in the *European Journal of Cancer* 1993, **29A**, 1958–1963. There was an error on p. 1960, in the legend to Table 2. The first sentence should read: *Figures in parentheses are the fraction of cultures showing growth.