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CASE REPORT

Proximal-Type Epithelioid Sarcoma in the Groin Presenting as a Diagnostic Dilemma

Abul Ala Syed Rifat Mannan · Amre A. Rifaat · Mirza Kahvic · Kusum Kapila · Mrinmay Mallik · Vinod Kumar Grover · Chandramouli Bharati · Arie Perry

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Abstract Epithelioid sarcoma is an uncommon soft-tissue sarcoma typically presenting as a subcutaneous or deep dermal mass in the distal extremities of young adults. Lately, a 'proximal' subtype has been described, which occurs in the pelvic and genital areas of somewhat older individuals and tends to behave more aggressively than the conventional subtype. The correct diagnosis of this subtype is essential, since this tumor can be easily mistaken for other malignant tumors that exhibit epithelioid morphology. We report a case of proximal-type epithelioid sarcoma that presented as an inguinal mass in a 47-year-old man. Histologically, the tumor consisted of diffuse sheets of epithelioid cells with scattered rhabdoid morphology. By immunohistochemistry, the neoplastic cells expressed cytokeratin, epithelial membrane antigen, vimentin, CD34, CD99 and showed complete loss of nuclear INI1 protein expression. Fluorescence *in situ* hybridization was considered borderline for 22q deletion. We present this case to emphasize the importance of diagnosing this uncommon tumor and the role of INI1 immunohistochemistry in establishing the diagnosis.

Keywords Epitheliod sarcoma · FISH · INI1 · Immunohistochemistry · Malignant rhabdoid tumor

Abbreviations

ES	Epithelioid sarcoma
CT	Computed tomography
MRI	Magnetic resonance imaging
Gd	Gadolinium
CEA	Carcinoembryonic antigen
FNA	Fine needle aspiration
EMA	Epithelial membrane antigen
CK	Cytokeratin
FISH	Fluorescence *in situ* hybridization
MRT	Malignant rhabdoid tumor
MPNST	Malignant peripheral nerve sheath tumor

Introduction

Proximal type epithelioid sarcoma was first described by Guillou et al [1] as an aggressive variant of epithelioid

A. A. S. R. Mannan · A. A. Rifaat · M. Kahvic
Department of Pathology, Al Jahra Hospital,
Jahra, Kuwait

K. Kapila
Department of Pathology, Faculty of Medicine,
Kuwait University,
Kuwait City, Kuwait

M. Mallik
Department of Cytopathology, Mubarak Al Kabir Hospital,
Jabriya, Kuwait

V. K. Grover
Department of Surgery, Al Jahra Hospital,
Jahra, Kuwait

C. Bharati
Department of Radiology, Al Jahra Hospital,
Jahra, Kuwait

A. Perry
Department of Pathology and Immunology,
Washington University School of Medicine,
St. Louis, MO, USA

A. A. S. R. Mannan (✉)
Department of Pathology,
Al Jahra Hospital,
P.O. Box 62276, Jahra 02153, Kuwait
e-mail: mannanrifat@rediffmail.com

sarcoma that differs from the conventional subtype by its propensity to occur in the pelvis, perineum and genital area of middle-aged or older adults. Histologically the tumor is characterized by diffuse proliferation of epithelioid cells with prominent 'rhabdoid' phenotype, which are indistinguishable from a host of other malignant tumors that exhibit 'epithelioid' cytomorphology [1]. We present this uncommon tumor in a 47-year-old man, emphasizing the role of immunohistochemical and molecular studies for INI1 status in establishing the diagnosis. The *INI1 (hSNF5/SMARCB1)* gene is a member of the SWI/SNF chromatin remodeling complex located on chromosome 22q11.2 [2]. It is characteristically deleted and/or mutated in malignant rhabdoid tumor (MRT), a tumor predominantly afflicting infants [3–5]. Immunohistochemical analysis to demonstrate the loss of INI1 protein expression has been found to be particularly helpful in establishing the diagnosis of MRT and its differentiation from composite rhabdoid tumors, the latter representing a wide variety of mostly adult type neoplasms that secondarily develop rhabdoid morphology, but usually retain INI1 expression [6–8]. Complicating this conceptual model however, several recent studies have reported that INI1 expression is characteristically lost in both conventional and proximal-type epithelioid sarcoma as well [9–12].

Case Report

A 47-year-old man presented with a gradually enlarging, painless soft tissue swelling of 1-year duration, in the right groin. Clinical examination revealed an oblong 10×7 cm, hard, mobile mass in the right inguinal region extending to scrotum. Both the testes, spermatic cords and the penis were normal. Computed tomography (CT) scan revealed a solid enhancing mass lesion in the right groin extending to the perineum and infiltrating the penile root. MRI showed the mass to be of moderate signal intensity, with strong enhancement after Gd-chelate administration (Fig. 1). Both spermatic cords were free and appeared normal. Bilateral testes and epididymes were normal. There was no evidence of lymphadenopathy anywhere. All the intraabdominal organs appeared normal. Chest X-ray also revealed normal study. Serum carcinoembryonic antigen (CEA) levels were normal; i.e. 1.8 ng/ml (normal range 0.3–2.7 ng/ml). A fine needle aspiration (FNA) of the nodular swelling was attempted. The smears were cellular with loosely cohesive tumor cells seen singly, in loose aggregates at places surrounding fibrovascular connective tissue (Fig. 2a).The cells were round to oval with moderate to abundant eosinophilic cytoplasm which appeared vacuolated in some cells. The nuclei showed variable pleomorphism, with vesicular chromatin and prominent nucleoli (Fig. 2b, c).

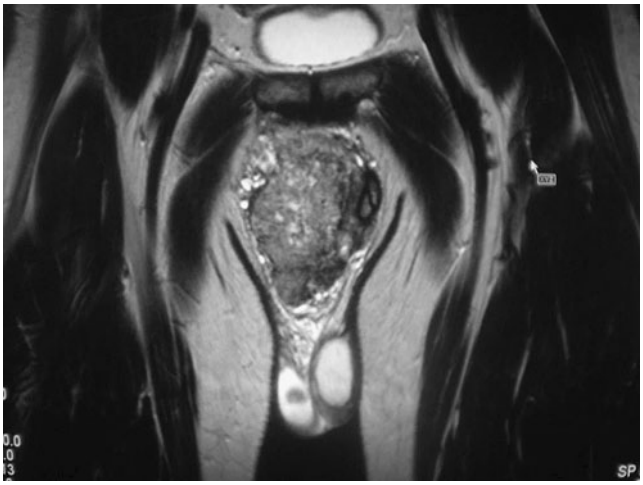

Fig. 1 T2 weighted coronal MRI image showing a mass in the perineum on the right side, depicting heterogeneous signal intensity. The mass is invading the right corpora cavernosa and displacing the shaft of penis to the left

Many binucleated and multinucleated cells with signet ring forms were seen. Mitoses were frequent. The tumor cells stained strongly positive for cytokeratin (Fig. 2d) and CD34 (Fig. 2e). A cytologic diagnosis of a poorly differentiated malignant tumor, possibly an epithelioid sarcoma was rendered. Histologic confirmation was advised.

On surgical exploration, the huge mass was extending from groin to the perineum. It was fused to fascia of perineal muscles and infiltrated the right crura. The mass was excised along with part of the crura. At frozen section, the surgical margins were free of tumor. On gross pathologic examination, the resected mass measured 14×7×2 cm. The tumor was well-circumscribed, firm and gray-white with focal areas of necrosis. On microscopy, it was similarly well-circumscribed and showed a multinodular growth pattern, composed of sheets of large epithelioid cells with vesicular nuclei, prominent nucleoli and abundant eosinophilic cytoplasm (Fig. 3a). At places cellular disintegration was observed, imparting a 'pseudoangiosarcomatous' pattern (Fig. 3b).There was moderate nuclear atypia. Mitotic figures were very infrequent (about 3 per 50 high power fields). Many cells had abundant clear cytoplasm with focal signet ring appearance (Fig. 3c). Occasional cells revealed 'rhabdoid' phenotype, characterized by intracytoplasmic, paranuclear globules. Interspersed were scattered, multinucleated osteoclast-like giant cells (Fig. 3d) and focal metaplastic bone formation. There were multiple foci of vascular invasion. Immunohistochemical stains were carried out with a wide panel of markers (Table 1). The epithelioid cells showed strong and diffuse immunoreactivity for pan-cytokeratin, epithelial membrane antigen (EMA) (Fig. 4a) and CD 34 (Fig. 4b). There was focal positivity for vimentin (Fig. 4c) and carcinoem-

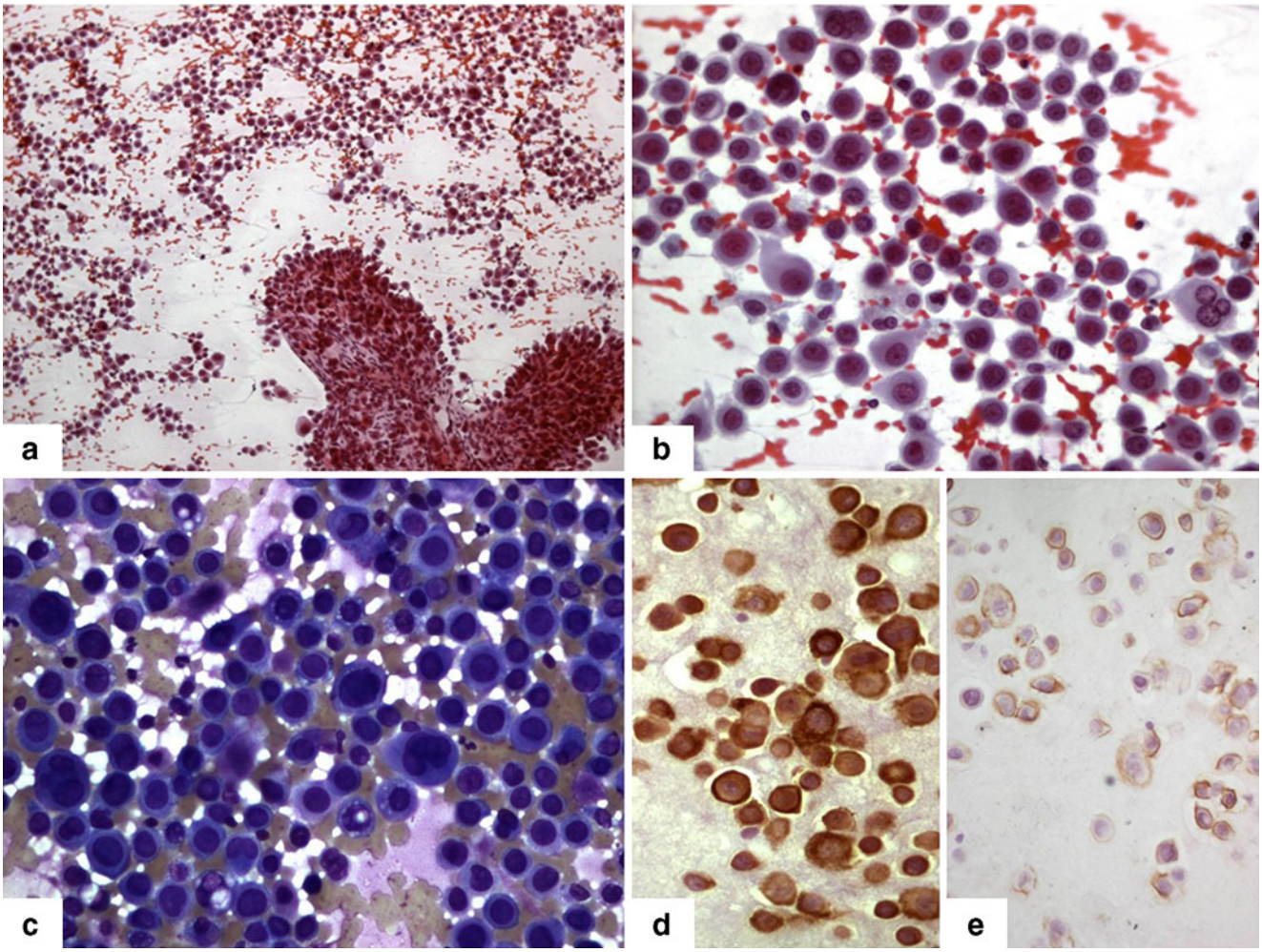

Fig. 2 Aspirate showing tumor cells singly and arranged around fibrovascular tissue (**a**); pleomorphic cells with moderate and vacuolated cytoplasm (**b**, **c**); tumor cells showing positivity for cytokeratin (**d**) and CD34 (**e**). (**a**: Papanicolaou stain × 200; **b**: papanicolaou stain × 400; **c**: May Grunwald Giemsa stain × 400; **d**, **e**: avidin biotin peroxidase × 400)

bryonic antigen (CEA). CD99 was also diffusely expressed (Fig. 4d). INI1 (BAF47) immunohistochemistry showed complete loss of expression in tumor cell nuclei with retained expression in endothelial cells, the latter providing an internal control (Fig. 5). A variety of other markers were also negative, including desmin, smooth muscle actin, Myo D1, calretinin, cytokeratin 5/6, S-100 protein, HMB-45, CD31,CD-117 and leukocyte common antigen.

FISH analysis was performed as previously reported using DNA probes for BCR on 22q11.2 (Spectrum Green-labeled**)** and NF2 on 22q12 (rhodamine-labeled) [8]. The commercially available BCR probe is located within 0.5 Mb of the INI1 gene and is therefore a useful surrogate marker for this gene at the DNA level. The results were borderline or equivocal for 22q deletion, as there were about 25% cells with only one signal, which was near the cut off for deletion (Fig. 6).Therefore, it was interpreted that these cells either did not have a deletion of this region or harbored it in only a small subset of tumor cells.

The patient had an uneventful post-operative period and was discharged on the fourth post-operative day. He is currently disease free, 14 months after surgery.

Discussion

In 1970, Dr. Enzinger first characterized epithelioid sarcoma (ES) as a distinct soft tissue neoplasm that typically occurs in the distal extremities of young adults [13]. Microscopically, most tumors show a characteristic granuloma-like pattern, with nodules of spindled and epithelioid cells surrounding areas of central necrosis. In 1997, Guillou et al [1], described a more aggressive subtype, termed 'proximal-type ES', which is commonly seen in the pelvis, perineum and genital area of middle-aged or older adults. A microscopically distinctive feature of the tumor is the predominance of large carcinoma-like 'epithelioid' cells, with frequent 'rhabdoid' morphology [1]. The tumor

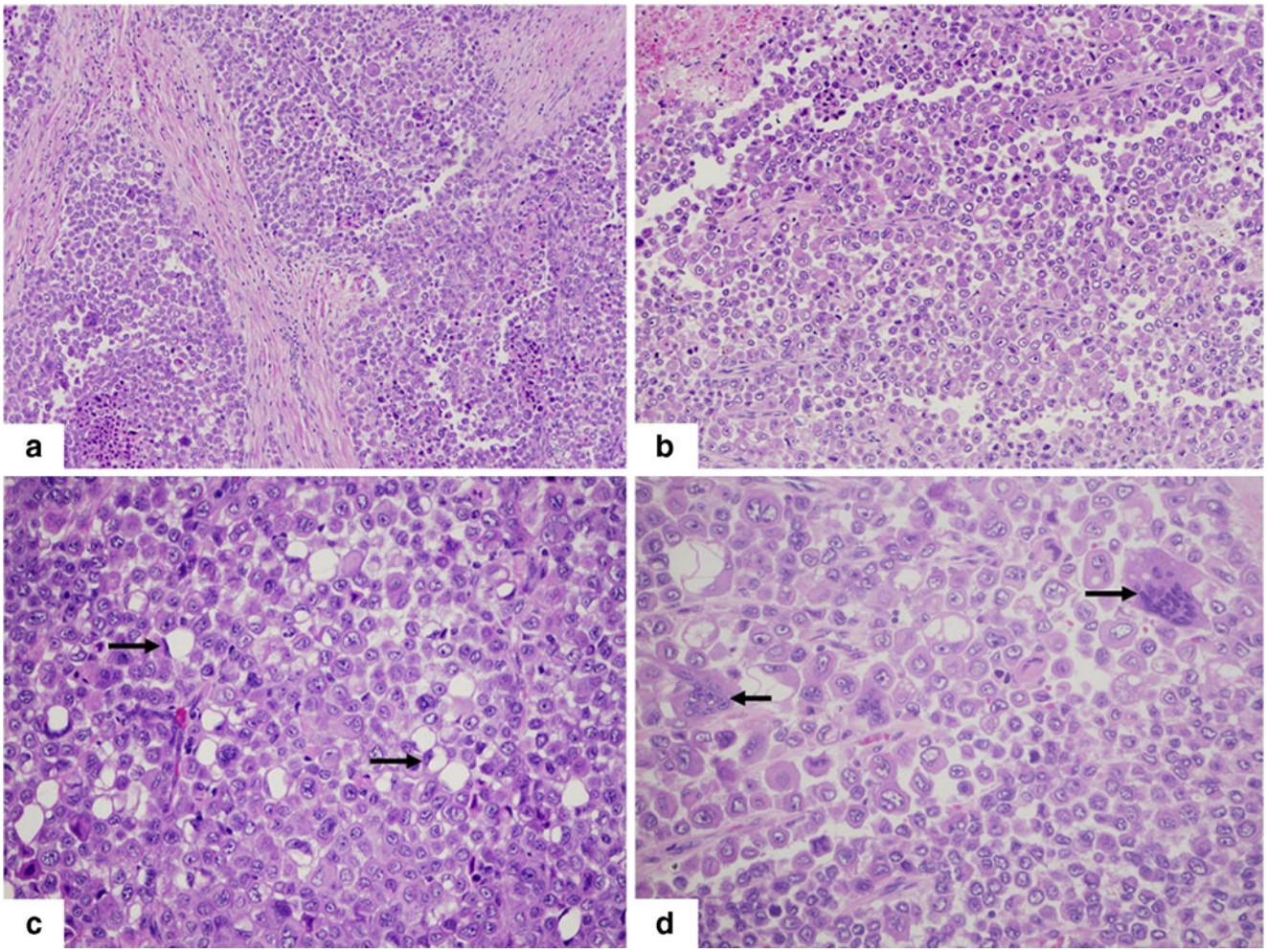

Fig. 3 Proximal type epithelioid sarcoma in the groin. **a** Nodular aggregates of tumor cells invested by fibrous septa; **b** Disintegrating tumor cells imparting 'pseudoangiosarcomatous' pattern; **c**, **d** Sheets of large polygonal cells with abundant eosinophilic cytoplasm admixed with many clear cells having 'signet ring' like morphology (arrow in **c**) and multinucleated osteoclast like giant cells (arrow in **d**). (**a**, **b**: hematoxylin & eosin × 200; **c**, **d**: hematoxylin & eosin × 400)

however retains the immunophenotypic profile of the classic ES, with co expression of cytokeratin (CK) and vimentin along with reactivity for CD 34 in about 50% of cases [1, 13, 14]. Recently, a loss of nuclear INI1 expression has been found to be a characteristic feature of both conventional and proximal-type epithelioid sarcomas [9–12].

The present case was identified in a middle-aged man as an inguinal mass. On morphology, the tumor showed sheets of large epithelioid cells with a prominence of clear cells with focal signet ring appearance. These features invoked a wide differential diagnosis that included metastasis from a poorly differentiated carcinoma, malignant rhabdoid tumor (MRT), epithelioid angiosarcoma, epithelioid sarcoma like hemangioendothelioma, epithelioid rhabdomyosarcoma, epithelioid malignant peripheral nerve sheath tumor (MPNST), mesothelioma and melanoma. A wide panel of immunohistochemical markers was performed to resolve this diagnostic dilemma as experienced by previous authors as well [1, 14].

The absence of another primary tumor on clinical and radiologic work up, a normal serum CEA level and CD 34 positivity in the tumor cells helped to exclude metastatic carcinoma. Variable reactivity for CEA has been described in epithelioid sarcoma, as was noted in the present case [15]. The tumor cells exhibited prominent epithelioid morphology on both cytologic and histologic sections, highly reminiscent of epithelioid mesothelioma. However, absence of reactivity for cytokeratin 5/6 and calretinin argued against this possibility. Lack of S-100 protein, HMB-45, desmin and smooth muscle actin expression ruled out other possibilities, such as epithelioid MPNST, malignant melanoma and myogenic tumors respectively. Epithelioid angiosarcoma was another strong differential

Table 1 List of various antibodies used

Antigen	Type	Dilution	Pretreatment	Source
Vimentin	V9	1:200	Microwave	Dako, Glostrup,Denmark
Desmin	D33	1:50	Microwave	Dako
SMA	My10	1:100	Microwave	Dako
Myo D-1	1A4	1:200	Microwave	Dako
Cytokeratin	AE1/3	1:100	Microwave	Dako
CK 5/6	Monoclonal	1:200	Microwave	Dako
EMA	E29	1:100	Microwave	Dako
CEA	Polyclonal	1:400	Microwave	Dako
S-100	Polyclonal	1:200	Microwave	Dako
HMB-45	Monoclonal	1:50	Microwave	Dako
CD34	Monoclonal	1:20	Microwave	Dako
CD 31	JC/70A	1:50	Microwave	Dako
LCA	Monoclonal	1:100	Microwave	Dako
CD99 (MIC2)	0–13	1:50	Microwave	Dako
CD-117	Polyclonal	1:40	Microwave	Dako
Calretinin	Monoclonal	1:100	Microwave	Dako
INI1(BAF47)	Monoclonal	1:50	Microwave	BD Transduction Labs, Franklin Lakes, NJ,USA

CK cytokeratin, *EMA* epithelial membrane antigen, *SMA* smooth muscle antigen, *CEA* carcinoembryonic antigen, *LCA* leukocyte common antigen

diagnostic consideration as it can exhibit a more solid growth pattern and may be positive for both cytokeratin and CD 34. The presence of 'pseudoangiosarcomatous pattern' in the present case further supported this consideration. This morphologic feature was also noted by several authors as a misleading pattern in cases of proximal-type epithelioid sarcoma [1, 14]. However, the lack of tumoral CD31 expression helped to exclude this possibility. In addition, diffuse expression of EMA was also helpful, since epithelioid angiosarcomas have limited to no reactivity for this marker [14]. The current case had a prominent component of clear cells with focal signet ring appearance that brought into consideration the possibility of epithelioid hemangioendothelioma, which could similarly be excluded by the lack of CD31 expression. Recently, 'epithelioid sarcoma-like hemangioendothelioma' has been described to show striking morphologic and immunophenotypic similarity to epithelioid sarcoma with co-expression of cytokeratin and vimentin [16]. But this tumor consistently expresses CD31 and is negative for CD34, whereas just the reverse is true for ES [14, 16], as was observed in the current case.

Loss of INI1 expression in the tumor cells was particularly helpful in establishing the correct diagnosis in the present case. Several recent studies have emphasized the utility of INI1 immunohistochemistry in confirming the diagnosis of ES by demonstrating loss of protein expression in the vast majority of both conventional and proximal subtypes [9–12]. Genomic deletion of INI1 has also been demonstrated in occasional cases of epithelioid sarcoma, most of which have been of the proximal type [9, 12, 17]. In the present case, FISH analysis was borderline for 22q deletion, further supporting the notion that most cases of ES do not have evidence of INI1 loss at the DNA level [12]. In terms of specificity, the vast majority of other sarcomas and epithelioid neoplasms retain INI1 expression [8, 10]. However, Hornick et al [10] recently reported loss of INI1 expression in 50% of epithelioid MPNST, while Perry et al [8] reported loss of INI1 expression in 1 case of retroperitoneal leiomyosarcoma. However, diffuse expression of S100 protein in the former and expression of myogenic markers in the latter should help in their differentiation from ES.

The distinction of epithelioid sarcoma from malignant rhabdoid tumor (MRT) needs special consideration, which is often difficult and riddled with controversy. Both tumors exhibit striking morphologic and immunophenotypic overlap, raising the question of their potential relationship. MRT is an extremely aggressive childhood malignancy characterized by the pathognomonic 'rhabdoid' cells with intracytoplasmic inclusions [18, 19]. These cells are present in various proportions in proximal type epithelioid sarcomas [1, 14], as was observed focally in the present case. Both tumors show similar polyphenotypic profiles with coexpression of epithelial markers, vimentin and CD99 [1, 14, 18, 19]. Loss of INI1 expression is also common to both MRT and epithelioid sarcoma, further compounding the diagnostic conundrum. Expression of CD34, as seen in the present case, helps to distinguish proximal-type epithelioid sarcoma (positive in 50% of cases) [1, 14] from MRT (consistently negative) [18, 19]. Recently, Kohashi et al

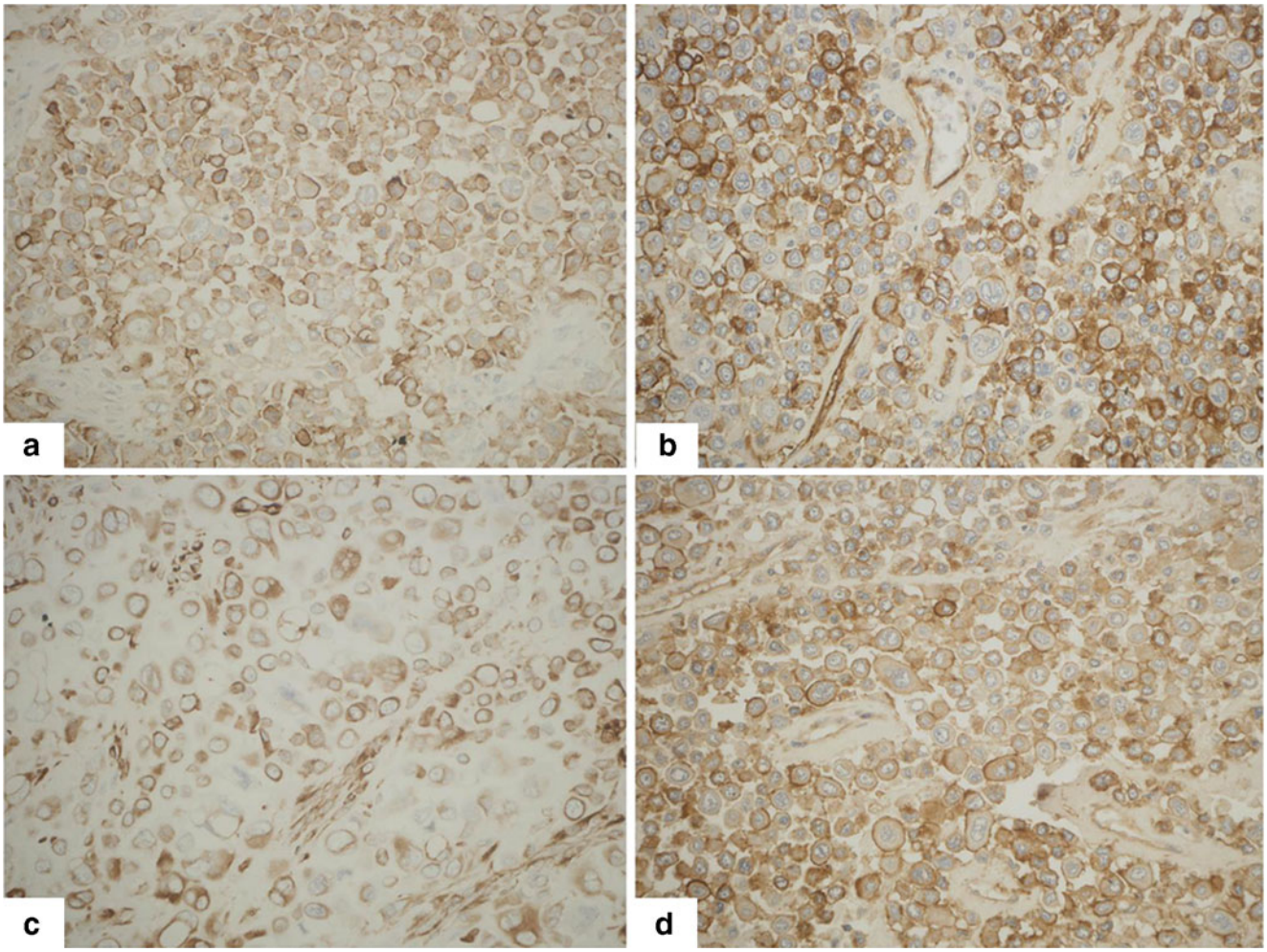

Fig. 4 Immunohistochemical profile. Tumor cells showing diffuse positivity for Epithelial membrane antigen (**a**), strong membranous positivity for CD 34 (**b**), focal positivity for vimentin (**c**) and diffuse positivity for CD 99 (**d**). (**a**, **b**, **c**, **d**: avidin biotin peroxidase × 400)

demonstrated differences at the genetic level between epithelioid sarcoma and MRT [12]. They observed that only in 10% of cases of ES with loss of INI1 protein expression had *SMARCB1/INI1* gene alterations at the DNA level. This was significantly lower than that observed in MRT (86%).The authors further suggested that the loss of SMARCB1/INI1 protein expression in ES may be associated with epigenetic or transcriptional level changes, but that in MRT, it may be caused by the gene alteration itself. This study thereby suggested that disregarding their morphologic and immunophenotypic overlap, ES and MRT are distinct tumor entities in respect to the mechanism of suppression of the SMARCB1/INI1 protein [12]. Hence, an analysis of *SMARCB1/INI1* gene alterations has the potential to become an important ancillary aid in the differential diagnosis of proximal-type ES and MRT.

The importance of identifying this subtype lays in its more aggressive clinical behavior than the classic type, with increased recurrence, early development of metastasis and a

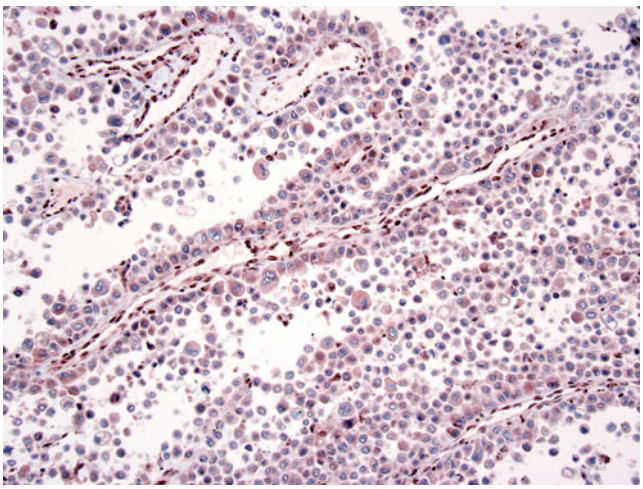

Fig. 5 Tumor cell nuclei showing complete loss of INI1 expresion. Note positive internal control in intratumoral endothelial cell nuclei (avidin biotin peroxidase × 200)

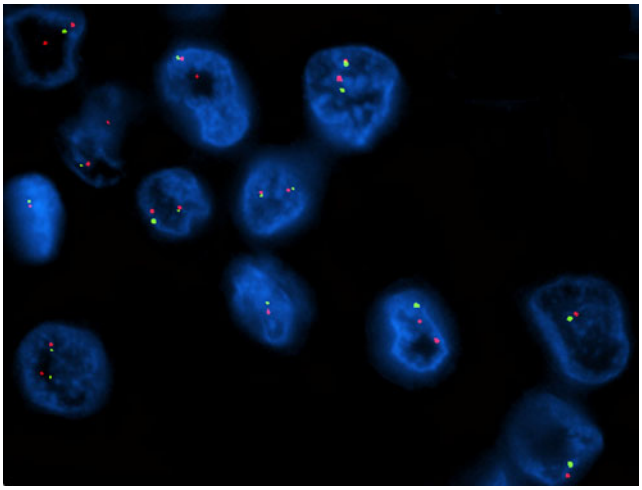

Fig. 6 FISH image showing borderline loss of 22q. A small subset of tumor cell nuclei show only one green (BCR) and one red (NF2) signal, while the remaining nuclei show normal 22q dosages with two green and two red signals. Given that these studies were performed on paraffin sections however, the possibility of nuclear "truncation artifact" from cutting thin sections cannot be entirely excluded. The 25% fraction of cells with loss of both signals is near the lab cutoff for deletions, suggesting that there is either no deletion or that only a small subset of tumor cells are deleted for 22q

higher incidence of tumor-related deaths. Hasegawa et al [14], in a study of 20 cases of proximal-type ES found that 65% of the patients developed local recurrence; the period between primary excision and first recurrence ranging from 1 month to 3 years and 8 months. Most patients had multiple recurrences. Seventy five percent developed metastases, primarily to the lymph nodes. Only 30% of the patients were alive at the last follow-up (2 months–9 years). In a recent comparative study between conventional ES (26 cases) and proximal-type ES (14 cases), Rekhi et al [20] found that the 7-year disease-free survival was 19.4% in the conventional ES and nil in the proximal subtype. The overall survival rate was also lower in the proximal ES (31.3%) than the conventional type (90.2%). Recurrences (83.3%) and metastases (41.6%) were also found to be more common in the proximal subtype.

The prognostic factors for this undifferentiated tumor are not yet clearly defined, as there have been very few small studies on this entity to date [14, 20]. In the study by Hasegawa et al [14], tumor size was found to be an adverse prognostic factor; survival of patients whose neoplasms measured 7.8 cm or more being significantly shorter than those with smaller tumors. However, they did not find significant prognostic implications for other parameters such as patient age and sex, tumor depth, histologic subtype, vascular invasion, tumor necrosis, mitotic count, p53 expression and MIB-1 index . Rekhi et al [20] observed that deeper location, larger tumor size (more than 5 cm) and higher tumor stage had an adverse affect on the outcome. It is debatable whether the presence of 'rhabdoid' phenotype per se connotes a worse prognosis for this tumor [14, 20, 21]. Further studies with fairly larger number of proximal-type ES are necessary to properly evaluate the prognostic impact of all these parameters.

Experience with the appropriate therapeutic approach for epithelioid sarcoma is also limited owing to the paucity of published reports in the literature. Wide surgical excision, with clear resection margins remains the primary goal [1, 14, 20]. In a recent study, Livi et al [22] investigated the role of postoperative radiotherapy in 22 patients with ES. Thirty percent of the patients developed local recurrence and half of them developed metastases despite further surgery, subsequently dying of disease. Others have however demonstrated better local control with adjuvant radiotherapy [23]. Onol et al reported partial response to adjuvant chemotherapy with adriamycin, ifosfamide and mesna in a case of metastatic proximal type epithelioid sarcoma of the scrotum [24].

In conclusion, the present case highlights the difficulty in diagnosing proximal-type epithelioid sarcoma as it has a bewildering variety of histologic mimics. Its aggressive behavior underscores the importance of a correct diagnosis, which can be achieved by judicious utilization of a wide panel of immunohistochemical markers that should ideally include study of INI1 status both by immunohistochemistry and by molecular methods. A timely diagnosis is essential for determining further treatment strategy and close follow-up to look for recurrences and distant metastases.

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