

Case report

Primary cutaneous extraskeletal Ewing sarcoma of the scalp

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Abstract

Primary cutaneous extraskeletal Ewing sarcoma (PCES) is an exceptionally rare variant of the Ewing sarcoma family of tumors, typically arising in the dermis or subcutis without osseous involvement. Due to its clinical mimicry of benign lesions and poorly defined dermoscopic features, early recognition can be challenging. To date, fewer than 20 well-documented cases have been reported in the literature. A 21-year-old male presented with a subcutaneous scalp mass with a reported 3-year clinical evolution. Dermatologic examination revealed a well-circumscribed vascularized lesion, and dermoscopy showed a polymorphous vascular pattern with arborizing and linear vessels, along with rainbow areas. Magnetic resonance imaging showed a non-invasive soft tissue mass. Histopathology confirmed a small round blue cell tumor with cluster of differentiation 99 (CD99) expression, and fluorescence in situ hybridization (FISH) analysis demonstrated Ewing sarcoma breakpoint region 1 (EWSR1) rearrangement, confirming PCES. The patient responded well to neoadjuvant chemotherapy and underwent surgical and radiotherapeutic management. This case is one of the very few reported instances of PCES with detailed dermatologic and dermoscopic documentation prior to oncologic intervention. It underscores the critical role of dermatologists in identifying rare soft tissue tumors. Prompt biopsy and molecular confirmation remain essential, and multidisciplinary care is key to improving outcomes in this rare malignancy.

Keywords: CD99, cutaneous sarcoma, dermoscopy, extraskeletal Ewing sarcoma, FISH

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Introduction

Extraskeletal Ewing sarcoma (EES) is a rare soft-tissue malignancy belonging to the Ewing sarcoma family of tumors (ESFT), which also includes osseous Ewing sarcoma and related small round cell sarcomas characterized by shared molecular alterations (1). ESFT is defined by recurrent chromosomal translocations involving the Ewing sarcoma breakpoint region 1 (EWSR1) gene, most commonly t(11;22)(q24;q12), which lead to the formation of the oncogenic Ewing sarcoma breakpoint region 1–Friend leukemia integration 1 (EWSR1–FLI1) gene fusion, which encodes an aberrant transcription factor driving tumorigenesis (1, 4). Although EES may arise in a wide range of anatomic locations, primary cutaneous extraskeletal Ewing sarcoma (PCES), originating in the dermis or subcutaneous tissue without underlying bone involvement, is an exceptionally rare presentation (5–10, 18, 19). Owing to its rarity and frequently deceptively benign-appearing clinical presentation, PCES poses significant diagnostic challenges and is often initially misdiagnosed as a benign cutaneous or adnexal lesion (5–10, 18). Early recognition and prompt histologic and molecular confirmation are essential because ESFT requires timely multidisciplinary management to optimize outcomes (11–13, 18, 19).

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Case report

We report the case of a 21-year-old male, with no significant past medical history, who presented with a subcutaneous tumor involving the left occipito-parieto-temporal region of the scalp. The lesion evolved progressively over approximately 3 years. According to the patient, he first noticed a nodule after a cranial trauma during a physical altercation, with no evidence supporting a causal relationship. Then the lesion increased steadily in size, prompting him to seek care.

He underwent a prior surgical excision at a private clinic. The resected specimen was described as a soft hemorrhagic mass, and it was submitted for histopathological examination. Histology showed a malignant tumor proliferation arranged in sheets, small nests, and trabeculae, composed of relatively uniform round cells with rounded nuclei, fine chromatin, amphophilic cytoplasm, and numerous mitotic figures, within a scant stroma containing fibrous septa with capillary structures. An initial immunohistochemical panel showed no expression of CD3, CD10, or cytokeratin, thereby excluding lymphoid and epithelial differentiation. The lesion was therefore classified as an undifferentiated malignant round cell tumor requiring further immunohistochemical characterization. However, no definitive diagnosis was communicated to the patient, and the lesion recurred locally within weeks.

The patient was referred to our dermatology department for further evaluation. On examination, the tumor appeared as a well-defined subcutaneous oval mass measuring approximately 6 cm in its largest axis. It was soft in consistency, non-pulsatile, non-tender, and freely mobile over deeper tissues. The overlying skin was intact and non-infiltrated, with prominent tortuous vessels visible on the surface and preservation of peripheral hair (Fig. 1). There was no associated pain, pruritus, bleeding, or ulceration. The surrounding skin appeared healthy.

Dermoscopy revealed a polymorphic vascular pattern, composed of linear, tortuous, and arborizing vessels, along with discrete rainbow areas (Fig. 2). This pattern raised clinical suspicion for a malignant vascular or adnexal tumor, including cutaneous angiosarcoma or adnexal carcinoma. Examination of regional lymph node basins (cervical, occipital, and postauricular) showed no palpable lymphadenopathy.

Magnetic resonance imaging (MRI) of the brain and scalp showed a well-demarcated highly cellular soft tissue mass located in the subcutaneous fat of the left occipital scalp. The lesion measured 40 × 21 × 28 mm, corresponding to the solid enhancing tumor component, and exhibited high signal intensity on T2-weighted sequences and low signal on T1, with heterogeneous enhancement following contrast administration. This radiologic measurement accounts for the intratumoral mass and excludes the surrounding soft-tissue and vascular components, explaining the difference with the larger clinical dimension (approximately 6 cm) assessed on physical examination. There was no evidence of calvarial invasion or intracranial extension.

Following local recurrence and referral to our center, the previously excised histological material (paraffin blocks and slides) was retrieved and underwent expert pathological review with an extended immunohistochemical and molecular workup. Histopathological reassessment confirmed a dense proliferation of small round blue cells arranged in sheets and nests, with finely dispersed chromatin, amphophilic cytoplasm, and frequent mitoses. The tumor cells showed diffuse strong membrane positivity for cluster of differentiation 99 (CD99) and cytokeratin, and they were negative for ERG, S100, NSE, desmin, chromogranin A, synaptophysin, CD34, CK7, and CK20 (Fig. 3). Fluorescence in situ hybridization (FISH) analysis confirmed a rearrangement of the EWSR1 gene in 100% of the nuclei examined, establishing the diagnosis of primary cutaneous extraskeletal Ewing sarcoma.

The extension workup, including total body computed tomography scan and bone scintigraphy, revealed no evidence of bone involvement, distant metastasis, or any primary bony lesion, confirming the primary cutaneous nature of the tumor.

The patient was referred to the national oncology center, where he received three cycles of neoadjuvant chemotherapy using the VDC/IE protocol (vincristine, doxorubicin, cyclophosphamide / ifosfamide, etoposide). Treatment response was excellent, with marked radiological and clinical regression, leaving a poorly defined residual infiltrate with coarse calcifications.

Following oncologic reassessment, complete surgical excision of the residual lesion was performed. The patient subsequently received consolidative chemotherapy, followed by adjuvant radiotherapy to sterilize

the tumor bed. Regular dermatological and oncologic follow-up has been maintained. At the most recent follow-up, approximately 48 months after surgical excision, the patient remains disease-free, with no evidence of local recurrence or systemic metastatic involvement.

Discussion

Ewing sarcoma of soft tissues, also referred to as EES, is a rare, aggressive, poorly differentiated malignant tumor that primarily affects children and young adults during the first 2 decades of life (11–13, 18, 19). Among its even rarer presentations is the primary cutaneous form of PCES, which arises directly in the dermis or subcutaneous tissue without bone involvement (5–10, 18, 19). Our case represents this exceptional variant, confirmed by complete imaging and scintigraphy workup that excluded osseous or visceral origin. This “rarity within a rarity” is underrepresented in the literature and poses significant diagnostic challenges (5–10, 18).

Clinically, PCES often mimics benign subcutaneous tumors due to its smooth surface, lack of ulceration, and absence of significant pain or systemic symptoms (5–10, 18). This deceptive presentation necessitates a high index of suspicion from dermatologists, particularly in young patients presenting with solitary scalp tumors. In our case, the lesion appeared as a 6 cm soft oval subcutaneous mass with intact overlying skin, traversed by prominent tortuous vessels and no regional lymphadenopathy—an appearance easily mistaken for adnexal tumors, vascular proliferations, or cystic lesions (5–10, 17). The prolonged clinical evolution observed in our patient requires careful interpretation. Although Ewing sarcoma is classically regarded as an aggressive small round cell malignancy, several reports of primary cutaneous extraskelatal Ewing sarcoma describe a misleadingly benign clinical course, often presenting as slowly enlarging, painless subcutaneous nodules, particularly in the scalp and head and neck region (5, 7, 8, 10, 18). Tavakkoli and Mueller emphasized that cutaneous Ewing sarcoma may be a clinically distinct presentation compared to osseous disease, with differences in tumor behavior and outcomes (16). In this context, the 3-year history in our patient does not contradict the diagnosis but rather illustrates a recognized diagnostic pitfall of PCES (18). Importantly, although the lesion was first noticed after cranial trauma according to patient history, no clinical, imaging, or histopathological evidence supports malignant transformation from a post-traumatic lesion (11–13, 18). Similar diagnostic delays have been reported in head and neck Ewing sarcoma due to nonspecific initial presentation and deceptively benign clinical appearance (12, 13). The dermatologic examination was therefore critical in guiding the diagnostic process. Attention to lesion borders, consistency, vascularity, and mobility over deeper planes is essential because rapid growth, asymmetry, and vascular prominence should raise suspicion, even in the absence of ulceration or systemic symptoms (17).

Dermoscopy, although rarely documented in cases of PCES, may provide additional relevant clues suggestive of malignancy (17). In our case, dermoscopy revealed a polymorphous vascular pattern, including arborizing and linear irregular vessels, as well as focal rainbow structures, typically described in highly vascularized tumors and in certain melanocytic and non-melanocytic malignancies (17). As highlighted by Apalla et al., dermoscopic features of cutaneous sarcomas remain poorly defined and largely non-specific; nevertheless, recurrent findings such as polymorphic vessels, milky-red areas, and structureless areas should alert the clinician to a possible malignant process and prompt urgent histopathological evaluation (17).

Given the extreme rarity of PCES, dermoscopic criteria specific to this entity cannot currently be defined, and the observations reported here should be regarded as hypothesis-generating rather than as a diagnostic pattern. In particular, the vascular polymorphism observed in this case cannot be considered specific for PCES and does not allow reliable dermoscopic differentiation from other aggressive cutaneous vascular or mesenchymal malignancies, including Kaposi sarcoma, dermatofibrosarcoma protuberans, or angiosarcoma (17). Likewise, rainbow-like structures observed under polarized dermoscopy are a non-specific optical phenomenon reported across a broad spectrum of vascular and non-vascular tumors and should be interpreted as a clue prompting biopsy rather than as a diagnostic feature of a defined histopathologic entity (17). Importantly, in the context of a stepwise diagnostic algorithm, the combination of vascular polymorphism and focal rainbow areas proved clinically valuable because it immediately shifted the working diagnosis away from common benign scalp nodules toward an aggressive vascular or mesenchymal tumor, thereby functioning as a critical triage step that prompted early biopsy and subsequent molecular confirmation

(17).

To our knowledge, this report is the first to provide a detailed description of dermoscopic findings in PCES, thus highlighting the emerging role of dermoscopy in the early clinical suspicion of rare cutaneous malignancies.

On this basis, the differential diagnosis of PCES is broad and includes a wide spectrum of benign and malignant cutaneous and subcutaneous tumors (17). The lesion may be misdiagnosed as an epidermoid or trichilemmal cyst, particularly in the early stages, when it is painless and slow growing. Other benign mimickers include pilomatricoma and lipoma, especially when presenting as soft and mobile nodules beneath intact skin. Vascular lesions such as lobular capillary hemangioma or even angiolipoma may be considered due to the visible surface vascularization. In more atypical presentations, adnexal tumors such as spiradenomas or hidradenomas are often suspected, especially when the tumor is located in the scalp or face. Malignant entities that should be considered include cutaneous metastases, angiosarcoma, Merkel cell carcinoma, and dermatofibrosarcoma protuberans, particularly when lesions are firm, fixed, or ulcerated. Rapid enlargement, irregular vascular patterns, and dermoscopic signs of malignancy should prompt early biopsy and histologic confirmation (17).

Histologically, EES belongs to the “small round blue cell tumor” family (4, 18). It is composed of uniform round to oval cells with finely dispersed chromatin, scant cytoplasm, and a high mitotic index (4, 18). It may show pseudo-rosetting, necrosis, or hemorrhage (4, 18). Importantly, the differential diagnosis includes Merkel cell carcinoma, which shows positivity for CK20 in a perinuclear dot-like pattern, and neuroendocrine markers such as chromogranin and synaptophysin; lymphoma, particularly diffuse large B-cell lymphoma, which expresses CD45, CD20, or CD3 depending on lineage; rhabdomyosarcoma, which can mimic EES but is desmin- and MyoD1-positive; small cell carcinoma, typically CK-positive and TTF-1 positive; metastatic neuroblastoma, especially in pediatric patients, often positive for NSE, synaptophysin, and PHOX2B; and poorly differentiated adnexal carcinomas, which may express cytokeratins but lack CD99 and EWSR1 rearrangement (4, 17, 18).

Immunohistochemical analysis is pivotal for narrowing the differential. CD99 is diffusely positive in over 90% of EES cases and shows strong membranous staining, but it is not entirely specific (17, 18). Additional markers, including cytokeratin, FLI1, and NKX2.2, may aid in diagnosis, although none are exclusive (17, 18). The final and most definitive tool is molecular testing, particularly the detection of EWSR1 gene rearrangements through FISH or reverse-transcription PCR (RT-PCR) (4). In our case, FISH demonstrated EWSR1 rearrangement in 100% of the nuclei analyzed, confirming the diagnosis of PCES.

Imaging, particularly MRI, remains the preferred modality for local assessment (1). It typically shows a well-circumscribed pseudo-encapsulated lesion with low T1 and high T2 signal intensity and heterogeneous contrast enhancement (1). Our patient’s MRI confirmed a non-infiltrative subcutaneous lesion with high cellularity and no involvement of the underlying calvarium or brain parenchyma.

Treatment of PCES follows protocols for osseous Ewing sarcoma, given the absence of specific guidelines for the cutaneous form (11, 18, 19). Multimodal therapy combining neoadjuvant chemotherapy (usually with VDC/IE), complete surgical resection with clear margins, and adjuvant radiotherapy remains the cornerstone of management (10–13, 18, 19). Our patient responded excellently to initial chemotherapy, followed by planned surgical excision and radiation therapy.

Available data suggest that superficial ESFT, including primary cutaneous cases, may have a more favorable outcome than deep-seated soft-tissue or osseous Ewing sarcoma; however, the available evidence remains limited by small sample sizes and heterogeneous follow-up (18). In a clinicopathological series of genetically confirmed superficial ESFT, Machado et al. reported a relatively favorable prognosis compared with deep-seated tumors, while emphasizing the need for larger cohorts and longer follow-up to draw definitive conclusions (18). In the present case, the patient remains disease-free approximately 48 months after complete surgical excision, consistent with the favorable outcomes reported in superficial ESFT (18).

In our review of the literature, only a limited number of well-documented cases of PCES have been reported to date, underscoring the exceptional rarity of this entity (Table 1).

This case underscores the importance of meticulous dermatologic examination and early use of dermoscopy in identifying rare malignant tumors presenting in the skin (17). In rapidly growing scalp lesions

in young adults, particularly with atypical vascular dermoscopic features, clinicians must maintain a broad differential diagnosis that includes aggressive mesenchymal neoplasms such as PCES (17). Multidisciplinary collaboration between dermatology, pathology, radiology, oncology and surgery is essential to optimize outcomes in these rare but impactful diagnoses (11–13).

Conclusions

Ewing sarcoma is an uncommon malignancy, particularly its extraskeletal cutaneous variant. In clinical practice, the possibility of Ewing sarcoma should be suspected when evaluating a subcutaneous vascular tumor, as in this case. Diagnosis rests on characteristic histopathologic features, supported by molecular confirmation of EWSR1 gene rearrangements. Although historically associated with poor outcomes, survival has improved markedly with multimodal therapy: multi-agent chemotherapy, wide surgical resection, and adjuvant radiotherapy. This case highlights the importance of early dermatologic recognition, thoughtful use of dermoscopy, and timely referral, which together optimize patient management and improve prognosis.

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Table 1. Full review of the literature summarizing all reported cases of primary cutaneous extraskeletal Ewing sarcoma (PCES), including the present case.

Reference	Location	Age/sex	Clinical features	Histology + FISH	Treatment	Patient evolution / outcome
Allam et al. (1999) (12)	Head and neck	Multiple	Variable presentations	CD99+, EWSR1+	Chemo + surgery + RT	Mixed outcomes
Fonseca et al. (2000) (14)	Head and neck	12/M	Large facial tumor	CD99+, EWSR1+	Chemo + surgery	Improved post-treatment
Patel et al. (2005) (4)	Scalp	17/F	Violaceous nodule with vascular surface	Small round cells, CD99+, EWSR1+	Chemo + surgery + RT	Complete remission post-treatment
Kalra et al. (2010) (6)	Chest wall	13/F	Subcutaneous chest wall mass	Round blue cells, CD99+, EWSR1+	Chemo + surgery	Good response, no relapse reported
Ballester Nortes et al. (2012) (22)	Back	29/F	Reddish non-ulcerated angiomatous tumor	CD99+, CK+, EWSR1-FLI1+	Wide surgical excision	No recurrence at 2-year follow-up
Yuste et al. (2015) (5)	Scalp	14/M	Rapidly enlarging nodule	CD99+, EWSR1+	Chemo + surgery + RT	No recurrence at 12 months
Olson et al. (2017) (13)	Head and neck	Multiple	Facial/head tumors	CD99+, EWSR1+	Chemo + surgery	Favorable response
Tavakkoli & Mueller (2018) (16)	Cutaneous + osseous	19/F	Skin + bone lesions	Mixed histology, EWSR1+	Chemo + surgery	Controlled disease
Haas et al. (2018) (8)	Scalp (congenital)	Neonate	Congenital scalp mass	CD99+, EWSR1+	Chemo + surgery	Alive and well, no recurrence
Al et al. (2019) (7)	Scalp	14/F	Scalp mass in child	CD99+, EWSR1+	Chemo + surgery	Alive and well, no recurrence
Toquica et al. (2020) (23)	Scalp (metastatic)	20/M	Multiple cutaneous nodules	CD99+, EWSR1+ (metastasis)	Chemo + surgery	Died of metastatic disease
Knight et al. (2022) (15)	Face	22/F	Firm facial mass	CD99+, EWSR1+	Chemo + surgery + RT	Favorable outcome post-surgery and RT
Hagihara et al. (2022) (21)	Skull (metastasis)	37/M	Delayed skull metastasis	CD99+, EWSR1+	Chemo + surgery + RT	Late recurrence after years
Wright et al. (2022) (1)	Multiple sites	Various	Extensive imaging review	CD99+, EWSR1+	Chemo + imaging	Review study (no outcome)
Regmi et al.	Scalp (during	25/F	Ulcerated scalp	CD99+,	Chemo +	Partial

(2023) (19)	pregnancy)		lesion	EWSR1+, high mitotic index	surgery	remission during pregnancy
Caltavitturo et al. (2023) (2)	Mediastinum	18/M	Deep thoracic mass	CD99+, EWSR1+	Chemo + surgery	Not specified
Almohaisen et al. (2023) (10)	Head/neck	10/M	Neck mass with compression	CD99+, EWSR1+	Chemo + surgery	Favorable evolution, under follow- up
Present case	Head	21/M	Rapidly enlarging subcutaneous scalp mass with tortuous surface vessels	CD99+, EWSR1+, CK+, FLI1-, other lineage markers negative	Chemo (VDC/IE) + surgery + RT	Excellent response post-chemo, surgery planned, under active follow-up

M = male, F = female, CK = cytokeratin, CD99 = cluster of differentiation 99, EWSR1 = Ewing sarcoma breakpoint region 1, FLI1 = Friend leukemia integration 1 transcription factor, RT = radiotherapy, Chemo = chemotherapy, VDC/IE = vincristine, doxorubicin, cyclophosphamide / ifosfamide, etoposide.



Figure 1. Clinical presentation. Clinical photos showing a well-circumscribed oval subcutaneous scalp mass, measuring approximately 6 cm in its greatest diameter, located in the left occipito-parieto-temporal region: A) anterolateral view demonstrating the prominent exophytic contour of the lesion, B) posterior view highlighting the well-defined margins and subcutaneous nature of the mass, C) oblique lateral view showing the smooth surface and absence of ulceration, D) close-up view emphasizing intact non-infiltrated overlying skin, with prominent tortuous superficial vessels and preservation of peripheral hair growth, E) superior view illustrating the dome-shaped configuration of the tumor, F) inferolateral view confirming the regular contours and lack of surrounding inflammatory changes.

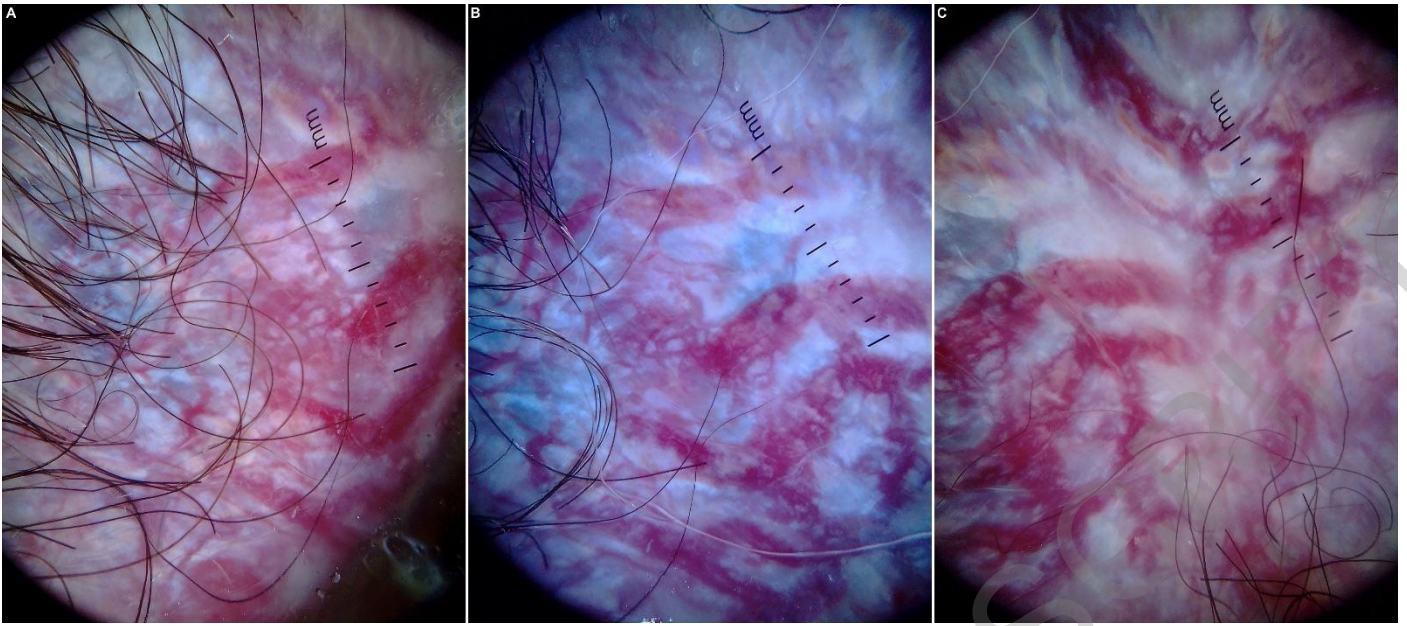


Figure 2. Dermoscopic features: A–C) polarized dermoscopy showing a polymorphous vascular pattern, characterized by a combination of linear irregular vessels, tortuous vessels, and arborizing vessels, set on a heterogeneous pinkish-white background. Focal rainbow areas are visible, reflecting an optical phenomenon related to complex tissue architecture and vascular organization under polarized light.

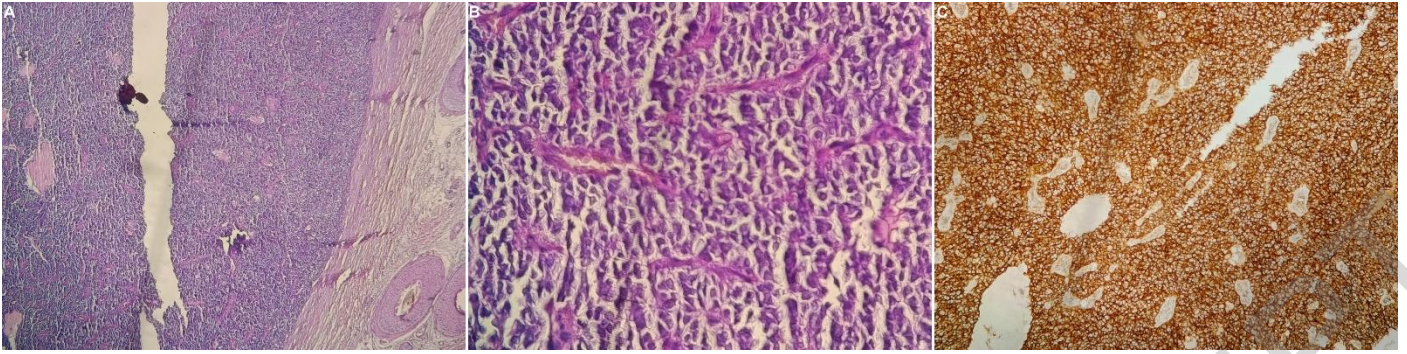


Figure 3. Histopathological and immunohistochemical findings: A) low-power view showing diffuse dermal and subcutaneous infiltration by a sheet-like malignant neoplastic proliferation, effacing normal cutaneous architecture (hematoxylin–eosin–safran, $\times 40$), B) high-power view demonstrating a dense monomorphic proliferation of small-to-intermediate-sized basophilic round cells (“small round blue cells”) with scant amphophilic cytoplasm, finely dispersed chromatin, inconspicuous nucleoli, and frequent mitotic figures (hematoxylin–eosin–safran, $\times 400$), C) immunohistochemical staining showing strong diffuse membranous expression of CD99 (MIC2) in tumor cells, supporting the diagnosis of Ewing sarcoma family of tumors.