RESEARCH PAPER

Primary superficial Ewing sarcoma with lymph node metastasis: Unusual presentation (*case report and review of literature*)

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Abstract

Primary superficial Ewing sarcoma (PSES) is an extremely uncommon tumor. Although Ewing sarcoma (ES) is primarily a bone tumor, it can sporadically affect the deep subcutaneous tissue or muscles, and it rarely occurs as a primary skin cancer. In contrast to bone or deep-seated Ewing sarcoma, superficial Ewing's sarcoma appears to have a better prognosis and rarely metastasizes. We report the case of a 21-year-old female who presented with a 1- year history of progressive enlarged right anterior thigh mass. The tumor was excised as a mass measuring 9×7×5 cm with a lymph node of 2×2 cm in size and sent for histopathology. Histopathological and immunohistochemical studies confirmed the diagnosis of PSES with lymph node metastasis, and the patient is now on treatment.

Key words: Superficial Ewing Sarcoma, lymph Node Metastasis

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Introduction

Ewing sarcoma is a primitive neuroectodermal tumor that affects bone, but it is unusual for it to present as a primary tumor in soft tissues including the skin.[1] Ewing sarcoma usually affects young males, in contrast with superficial Ewing sarcoma (SES) that occurs in older adolescent females.[2] The lower and upper extremities are the common sites of involvement, followed by the head, and trunk.[3] Primary superficial ES often manifests as a superficial solitary mobile oval mass, which is sometimes painful.[4] When compared to intra-osseous Ewing's sarcoma, primary superficial Ewing's sarcoma has a more favorable prognosis and a high survival rate.[2] Histologically, the tumor shows malignant small round cell tumor morphology with hyperchromatic nuclei and clear cytoplasm. Since primary superficial (cutaneous) Ewing sarcoma shares morphological similarities with other cutaneous round cell tumors, it is at risk of being misdiagnosed.[3] The diagnosis of PSES can be obtained with the use of immunohistochemistry (IHC), and other techniques such as cytogenetic and molecular genetics.[5] Surgical resection is the mainstay of treatment, followed by chemotherapy with or without radiotherapy.[3] Only a few cases of PSES with lymph node metastasis are documented in the literature.

Case report

A twenty-one-year-old lady presented with a history of a progressive enlarged palpable right anterior thigh mass for 1 year duration, which was completely resected. The past medical and family histories were unremarkable. Magnetic resonance imaging (MRI) of the right thigh revealed an 8.7 ×4.1 ×6.8 cm and 2.8 ×2.5 cm two well-defined anterior thigh mass. The lesions appear to have low signal intensity in T1 images and heterogeneous signal intensity with multiple cystic areas in T2 images **(Figure-1)**.

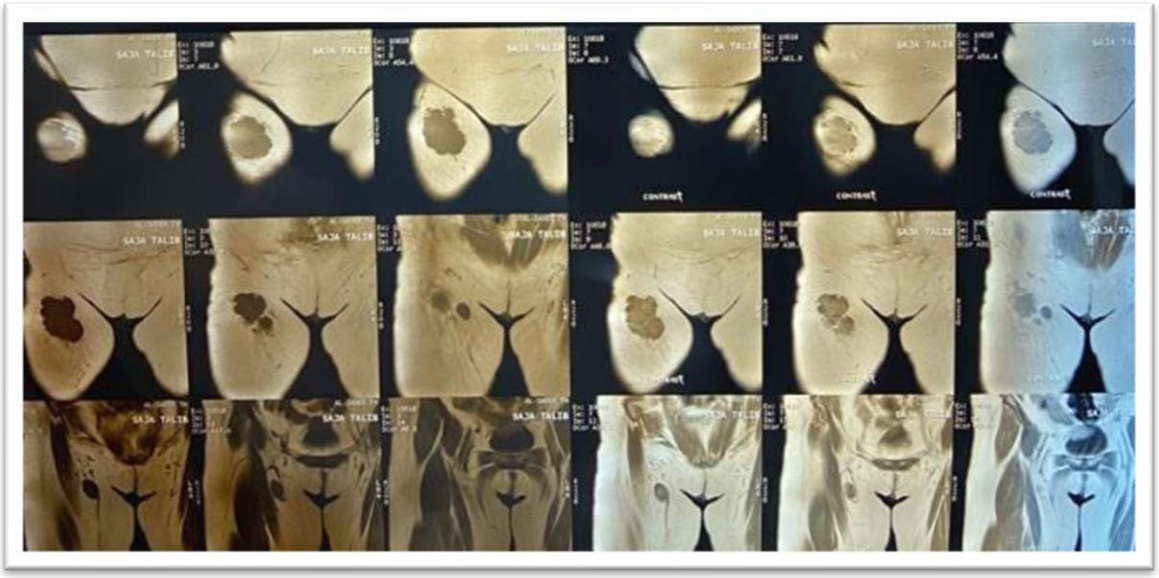

Figure 1: T 1- weighted magnetic resonance imaging (MRI) shows a low- intensity mass confined to the subcutaneous area and extending to cutaneous tissue. A: T 1- weighted images without contrast B: with contrast

A subsequent contrast-enhanced computed tomography (CECT) of the chest and abdomen was performed, and this revealed a few mesenteric lymph node enlargements in the mid abdomen, the largest one measures 1.2 ×0.8 cm. The patient underwent a wide local excision of the right anterior thigh mass with a single regional lymph node. Gross examination revealed a 9×7× 5 cm lobulated soft gray mass with skin and subcutaneous tissue in addition to a single lymph node of 2×2 cm in size. Histological examination revealed features of a subcutaneous multinodular tumor separated by thick fibrous septa. Each nodule consists of cells that have small undifferentiated round cell tumor morphology, with a few areas exhibiting Homer-Wright pseudorosettes. Moderate mitosis, in addition to areas of necrosis were presented **(Figures 2, 3, 4)**.

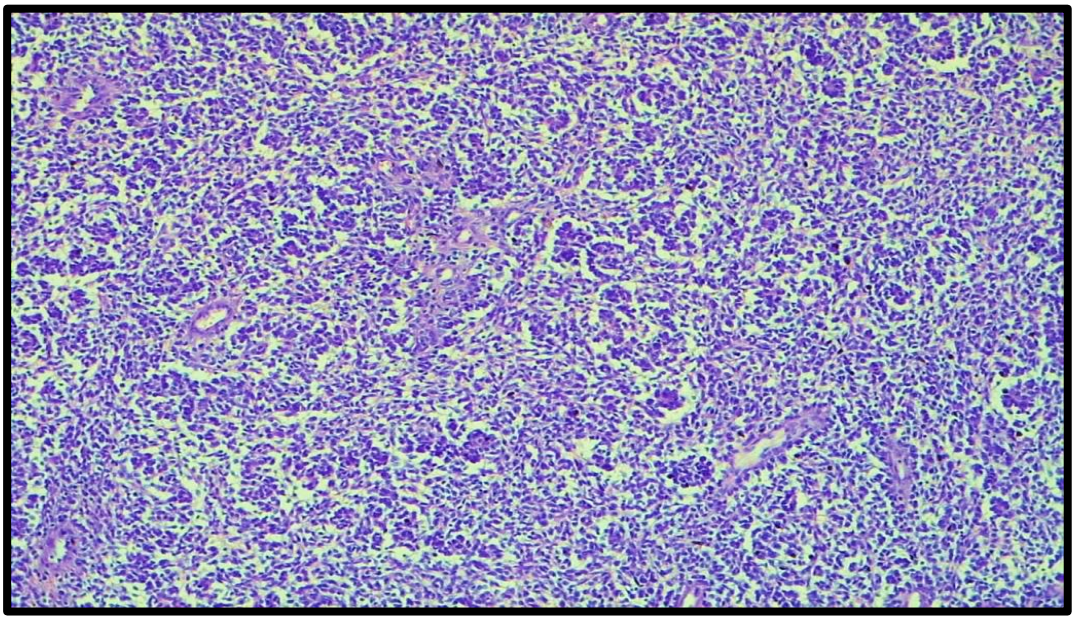

Fig 2. Primary superficial Ewing sarcoma, showing diffuse proliferation of small round blue cells (H&E, ×100).

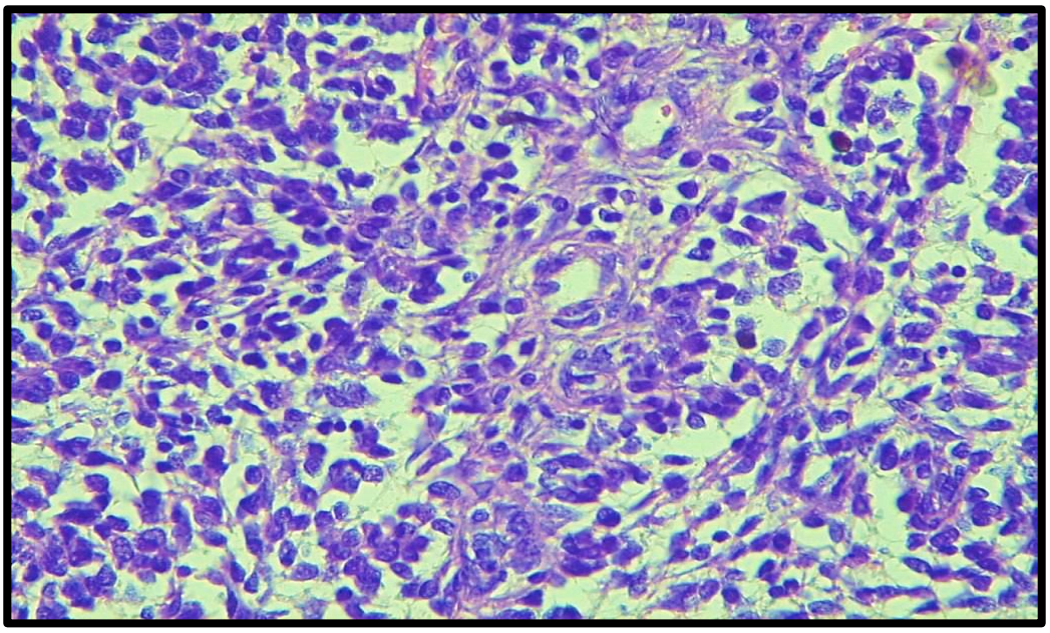

Fig 3. Primary superficial Ewing sarcoma, showing diffuse proliferation of round to oval cells with small basophilic nuclei and indistinct cytoplasm (H&E, ×400).

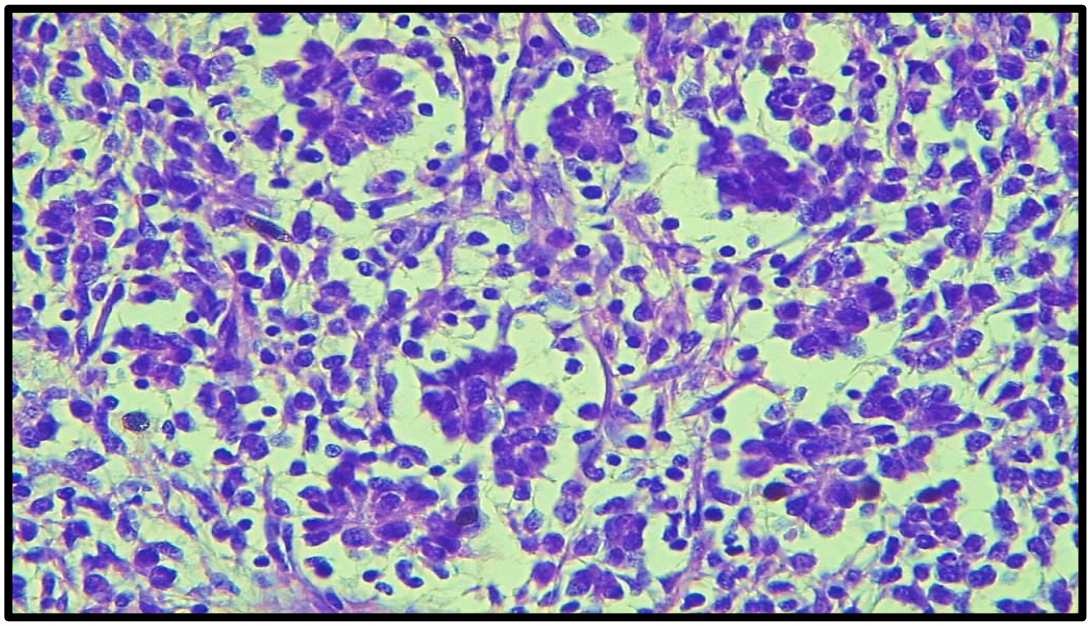

Fig 4. Primary superficial Ewing sarcoma, showing area of pseudorossettes (H&E, ×400).

A regional lymph node revealed a metastatic tumor **(Figure-5)**. The entire tumor was removed with a safe margin. By immunohistochemistry, the tumor cells were positive for vimentin, S100P, CD99 (which is typically strong positive in 90 - 95% of ES)[6], FLI-1 (which usually exhibits positive expression in 90% of tumors)[6], and Ki-67 > 29%, but negative for CK5/6, EMA, and GCDFP-15. The tumor cells were also negative for CK20, CD45LCA, Myogenin, Melan A and WT1 (these markers exclude Merkle cell carcinoma, lymphoma, rhabdomyosarcoma, malignant melanoma, and desmoplastic small round cell tumor, respectively)[6] **(Figures- 6, 7, 8, 9)**. The patient is currently on chemotherapy.

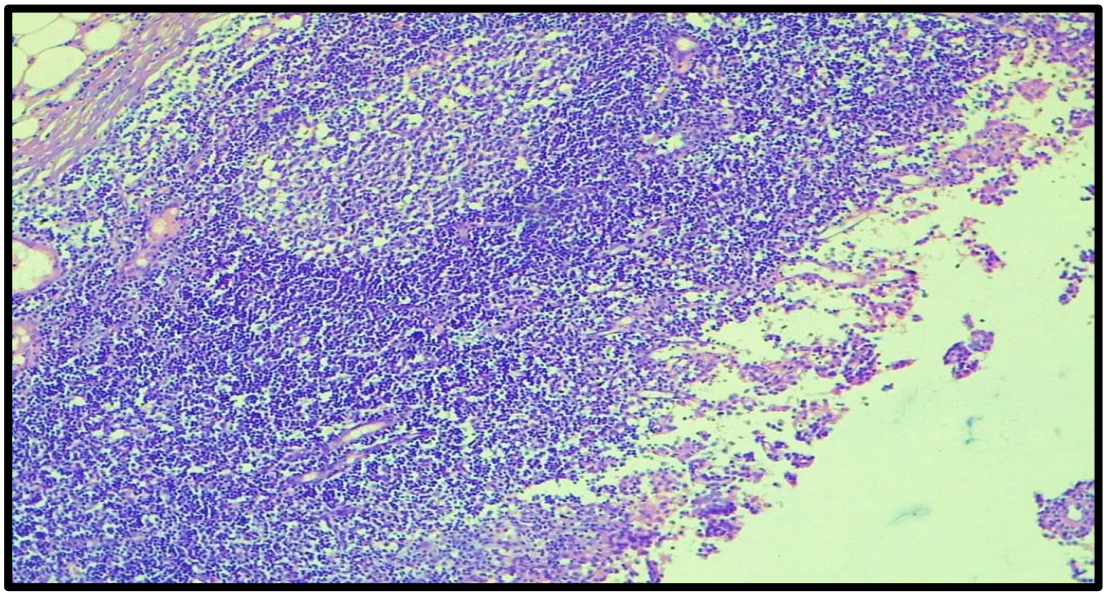

Fig 5 . Lymph node biopsy, showing nodal infiltration by metastatic malignant small round cells (H&E, ×100).

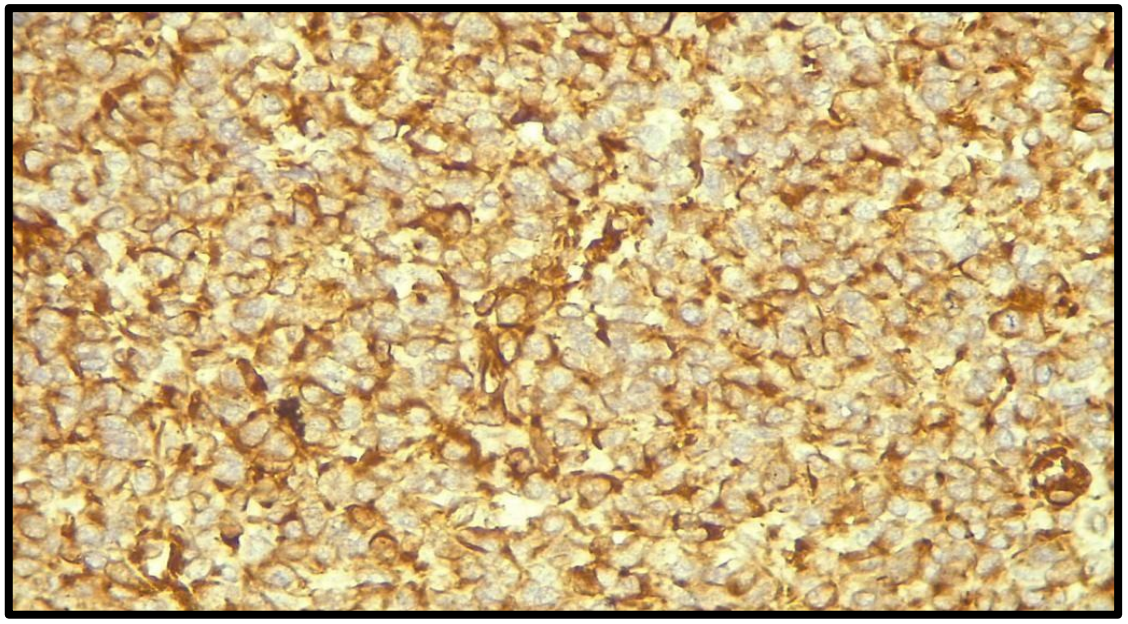

Fig 6. Vimentin positive expression in PSES (IHC,× 400).

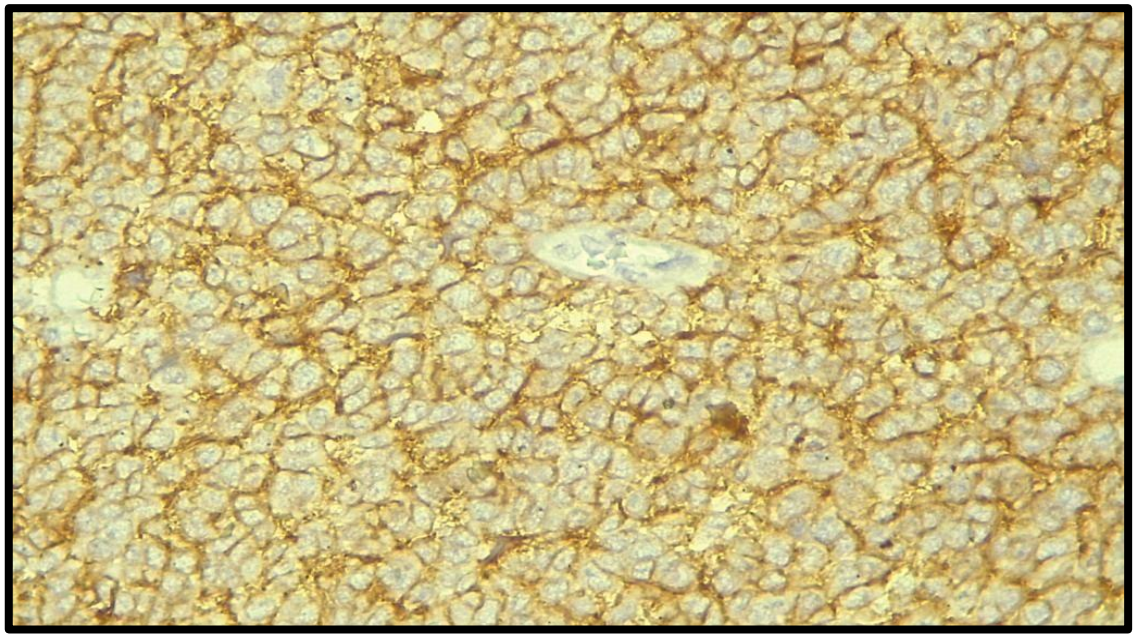

Fig 7. CD99 positive expression in PSES (IHC,×400).

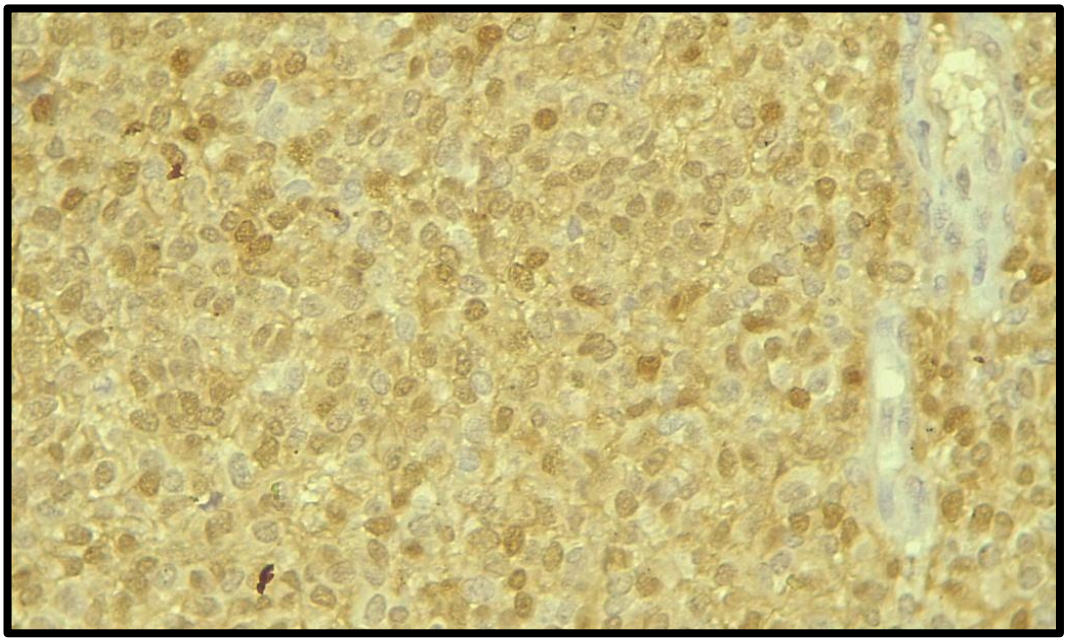

Fig 8. S100p positive expression in PSES (IHC, ×400).

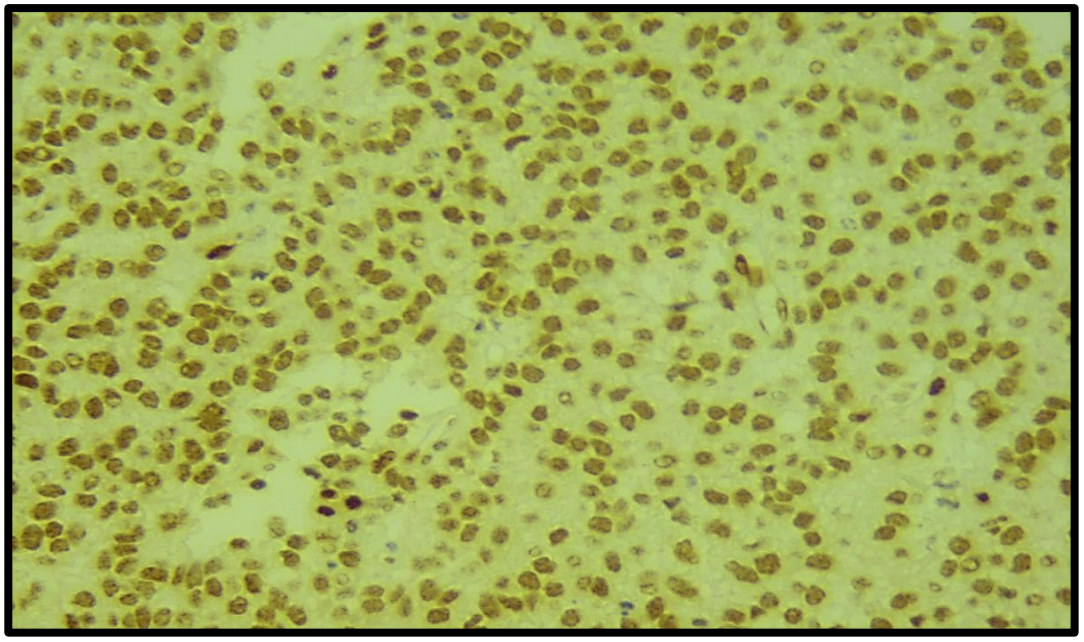

Fig 9. FLI-1 positive expression in PSES (IHC, ×400).

Discussion

A primary superficial Ewing sarcoma of the skin was first reported in 1975. It is a rare malignant round-cell tumor that mostly expresses CD99, which is also known as cutaneous or dermal ES, [7] and has a slightly delayed onset of presentation compared with skeletal ES.[7, 8] In contrast to our case, which is presented with lymph node metastases, the PSES was less aggressive than its deep soft tissue and bony counterparts, with an apparently more favorable prognosis.[9] A primary superficial (cutaneous) ES is very unusual. Only a few cases are reported in the literature. Previous studies reported that the lower limbs are the most commonly affected sites, with a female predominance for PSES (cutaneous ES), (**Table-1)**.

Primary Superficial Ewing Sarcoma with lymph node metastasis: unusual presentation (Case report and review of literatures)

Table 1. Literature review of 11 cases of primary superficial Ewing sarcoma

Case	year	Age (years)	Sex	Duration	Tumor site	Tumor size(cm)	Positive or negative for metastasis
1	2011[10]	17	Female	Several years	Mid back	5 × 6	Negative
2	2011[10]	12	Female	3months	Left thigh	4 × 8.5	Negative
3	2014[1]	26	Female	6months	Right clavicular region	3 × 4	Negative
4	2014[7]	9	Female	1month	Upper left back	1.5× 2	Negative in initial biopsy. Positive in recurrent tumor
5	2015[11]	37	Male	3 months	First toe of the left foot	9.5 × 8	Negative
6	2015[12]	13	Male	6 weeks	Sole of the right foot	1.5 × 0.6	Negative
7	2017[13]	10	Male	12 months	Left side chest wall	1.5 × 1.8× 1.3	Negative
8	2019[14]	17	Female	12 months	Right hand	0.5 cm in diameter	Negative
9	2020[2]	17	Female	4 years	Anterior left thigh	2.5 × 2.4 × 1.5	Negative
10	2021[15]	35	Female	2 years	Left posterior thigh	20 × 15	Negative
11	2022[8]	36	Female	3 years	Abdomen	2.5 ×1.5	Negative
Our case	2022	21	Female	1 year	Anterior right thigh	9 × 7 × 5	Positive

Conclusion

Although PSES (cutaneous ES) is quite rare, it should be taken into consideration while making a diagnosis of cutaneous round cell tumor and after excluding secondary involvement of the skin by skeletal ES. For individuals with PSES, adequate surgical resection is advised along with prompt evaluation and appropriate therapy.

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ورم إيوينغ السطحي الأولي المنتشر الى العقد الليمفاوية: (تسجيل حالة مرضيه ومراجعة للبحوث)

يعتبر ورم ايوينغ الاولي السطحي (PSES) من الاورام الغير شائعه. على الرغم من أن ورم إيوينغ (ES) هو في الأساس ورم يصيب العظم، إلا أنه ممكن ان يصيب في حالات معينه الانسجه الرخوه تحت الجلد و العضلات ، ونادرًا ما يحدث كسرطان جلدي أولي. على عكس ورم إوينغ في العظام و الانسجة العميقه، وجد ان ورم إيوينغ في الانسجه الرخوه والسطحية يكون افضل استجابة وقلما ينتشر. تم تسجيل حالة لامرأة تبلغ من العمر ٢١ عامًا تعاني من وجود كتله في الفخذ الايمن والتي ازداد حجمها تدريجيا خلال سنه كاملة. تم استئصال الورم ككتلة ذات ابعاد ٩× ٧ × ٥ سم مع عقدة ليمفاوية بحجم ٢ × ٢ سم وإرسالها إلى الفحص النسيجي. أكدت الفحوص النسيجية المرضية والكيميائية المناعية تشخيص ورم إيونك ((PSES مع انتشاره في العقدة الليمفاوية، والمريض الآن في مرحلة العلاج.

الكلمات المفتاحية: سوبيرفاسيال إيوينغ ساركوما, ليمف نود ميتاستاسيس