Rıb-Derived Lesion Mımicking Primary Pulmonary Ewing Syndrome

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ABSTRACT

Only sporadic cases of primary pulmonary Ewing sarcoma (PES) have been reported in the scientific literature. Only 4 of the 50 reported cases are in the pediatric age group. It should be monitored that these cases are truly PES. Advanced imaging should investigate whether the actual origin is another structure. Our case, which appeared to be PES, but was understood to have rib origin after advanced imaging (positron emission tomography-PET/CT) and the first course of chemotherapy. Our case was thought to be PES based on initial clinical imaging. However, after advanced imaging (positron emission tomography-PET/CT) and the first chemotherapy course, it was understood that it originated from the ribs. The diagnosis of PES is very rare and it is important for those diagnosed to be followed closely for the correct origin. We wanted to report this to emphasize the effect of appropriate imaging and follow-up on treatment success. We wanted to emphasize the impact of appropriate imaging and actual diagnosis on the type of treatment and its success.

CASE REPORT

INTRODUCTION

Ewing sarcoma (ES) is a rare neuroectodermal malignant neoplasm. It constitutes 6-8% of primary malignant bone tumors. It usually involves the long bones and pelvis [1]. While the majority of ES cases occur in children and bones, approximately 25% arise from extraskeletal (EES) [2]. EESs are relatively rare, accounting for approximately 1% of soft tissue sarcomas. Primary pulmonary ES (PES), which is EES, is rare. It has mostly been reported as a single case in the scientific literature. Approximately 50 cases have been published since 1989 [3]. Only 4 of the reported cases are in the children's age group (9 years old, 12 years old, 15 years old, 16 years old) [4-7]. The diagnosis of PES is very rare and it is important for those diagnosed to be followed closely for the correct origin. Our case, which appeared to be PES, was understood to have rib origin only after advanced imaging (positron emission tomography-PET/CT) and the first chemotherapy course. We wanted to emphasize the impact of appropriate imaging and actual diagnosis on the type of treatment and its success.

CASE REPORT

A 3.5-year-old girl patient with normal development was admitted to our clinic with the diagnosis of PES (Figure 1) as a result of the biopsy taken from the mass in the left lung. The patient had no other findings on physical examination and her complete blood count was normal; ESR 62 mm/h, CRP 13.2 mg/L, LDH 250 U/L, uric acid 5.4 mg/dL, ferritin 46.7 ml/ng, vitamin B12 322 pg/mL, NSE 23.55 ng/ml (increased), beta HCG 0 mIU/ml, alpha fetoprotein 0.2 ng/mL, and urine HVA 2.13 mg/24h, VMA 2.85 mg/24h. Bone marrow aspiration and biopsy were normal. In PET/CT, intense FDG uptake (SUVmax: 7.62) was observed in the 100X81X108 mm mass lesion, extending from the anterior mediastinum to the anterior of the upper lobe of the left lung, filling the upper lobe, narrowing the adjacent bronchus, and containing heterogeneous-looking necrotic areas. The mass was destroying the left 2nd rib posterior and extending towards the lung. The mediastinum was observed to have shifted to the right and the mass had erased the borders of the mediastinum in some areas. FDG uptake was present in an 8 mm sized nodular lesion (SUVmax: 2.06) located subpleurally in the apical part of the upper lobe of the right lung, and a 13X10 mm sized nodular lesion (SUVmax: 2.52) located paramediastinally in the middle lobe mediobasal, but a few millimetric sized nodules with no detectable FDG uptake were also observed (right lung metastatic nodules in the lower and upper lobes). A soft tissue dense lesion that was compatible with the thymus and did not show significant FDG uptake was selected in the anterior mediastinum, and a thin fatty plane was selected between it and the mass lesion observed in the left lobe. Mild hypermetabolic diffuse involvement (SUVmax: 1.63) in the splenic parenchyma was evaluated due to the inflammatory response (Figure 2).

Current standard chemotherapy includes vincristine, doxorubicin, and cyclophosphamide (VDC) alternating with IE for 12 weeks (six cycles) of induction given every 2 weeks: vincristine 1.5 mg/m^2 (maximal dose 2 mg) and cyclophosphamide 1200 mg/m^2 for 1 day, doxorubicin 37.5 mg/m^2/day for 2 days; ifosfamide 1,800 mg/m^2/day for 5 days and etoposide 100 mg/m^2/day for 5 days [8]. The CT after the 1st course of VDC (Figure 3) shows intermediate planes with mediastinum and lung, filling the middle-upper parts of the left hemithorax, measuring 71X93X89 mm at its widest point, containing well-circumscribed, necrotic areas, thought to originate from the left 3rd rib posterior. An intact, well-circumscribed, slightly heterogeneous enhancing solid lesion was observed. There was heterogeneity and cortical irregularity in the posterior left 3rd rib. Consolidation was observed in the upper lobe lingual segment due to lung compression. Several nodules in the right lung, the largest of which was 5 mm in diameter in the upper lobe posterior segment, were evaluated as metastases.

DISCUSSION

Etiology & Demographics

The most common primary bone locations for ES are lower extremity (45.6%), femur (20.8%), pelvic (20%), upper extremity (12.9%), axial skeleton/ribs (12.9%), ilium (12.5%), fibula (12.2%), tibia-humerus (10.6%), sacrum-ischium (3.3%), face (2.3%), feet-forearm (2%), pubis (1.7%) and hand (0.3%). Approximately 10-30% of patients have detectable metastases at the time of diagnosis. The most common metastatic sites are lung (38%), bone (31%) and bone marrow (11%). The location of the primary tumor is related to the incidence of metastasis at the time of diagnosis [8]. It was understood that our patient did not have PES after PET imaging and the first course of treatment. Surgery would be performed primarily in the diagnosis of PES. However, our patient's rib metastasis to the lung made surgery unnecessary. No involvement was detected in the hilar lymph nodes. There were 2 metastasis foci in the opposite lung. Surgery was prevented from unnecessarily delaying chemotherapy at the beginning of treatment. When the metastasis foci disappeared in PET repeated after chemotherapy, the mass, which had shrunk considerably, was removed with surgery. One year after completion of chemotherapy, our patient continues to live a healthy life.

Clinical & Imaging Findings

The general imaging properties of EESs are nonspecific. The majority present as a large and bulky soft tissue mass. Thoracopulmonary EES is rare in the pediatric group. It is even rarer in adults [2]. PES is a rare malignancy with only sporadic cases reported in the scientific literature [1]. The lung is a rare organ of primary involvement of EES, but is the most common site of metastasis [3]. Most cases present as a solitary, well-circumscribed heterogeneous mass or nodule with areas of low attenuation consistent with necrosis and relative hypoconcentration on contrast-enhanced CT [9].

The majority of thoracopulmonary EES occurs in white patients and males (60%). The average age at diagnosis is 30.5 years, with only a few cases diagnosed after age 50. The typical finding of thoracopulmonary EES is a painful chest wall mass with symptoms of cough, respiratory distress, weight loss, anorexia, Horner syndrome, or regional lymphadenopathy. The clinical presentation of the disease is generally severe, because of the tendency to grow rapidly and invade thoracic organs; therefore, only a small number of cases are resectable at presentation. In most cases, large tumors are observed, often invading the pleura (with or without pleural effusion) or the central airways and mediastinum. Severe clinical situations characterized by diffuse parenchymal invasion, massive pleural effusions, airway obstruction, and mediastinal shift, are frequent. Radiologically, thoracopulmonary EES often manifests as an aggressive unilateral pleural-based mass. Direct invasion of the chest wall musculature or ribs, mediastinum, or lung is common. Pleural effusions are often large, with loculated fluid sometimes forming a pseudotumor. Ipsilateral mediastinal or hilar adenopathy manifests in about 25% of thoracopulmonary EES cases. In our case, as in the majority, ipsilateral and hilar adenopathy was not detected. Calcifications occur in approximately 10% of cases. MRI may be particularly useful in helping to determine the presence and extent of chest wall muscle invasion [1, 2, 9]. In our case, PET/CT showed more direction regarding the primary location of the lesion.

The most specific feature uniting the vast majority of ES family of tumors (ESFT) cases is the aberrant fusion of the ES gene (EWSR1) with one or more transcription factors. A chromosomal translocation involving the long arms of chromosomes 11 and 22 (t[11;22] [q24;q12]) is the most common translocation, resulting in EWSR1-FLI1 fusion protein (85%). The second common translocation is t(21:22)(q22:q12), which results in EWSR1-ERG fusion (10%) [2].

The staging workup for EES includes an imaging evaluation for metastatic disease. Lung metastasis is by far the most common (27%). Therefore, chest CT is usually performed alone or in conjunction with PET owing to its superiority in detection of subcentimeter pulmonary metastases over MRI or PET alone. In our patient, CT and MRI could not provide sufficient guidance regarding the determination of the primary site. However, more information has been obtained with PET. Bone marrow aspiration and bone scintigraphy are performed to detect metastases in the skeleton. Both of them have been replaced by FDG PET/CT, which has higher sensitivity in detecting bone metastases [5]. PET also has the additional benefit of evaluating the primary tumor site and distant soft tissue metastases. Whole-body MRI may also play a role in staging but is performed less frequently, partly due to long acquisition times and limited availability [2].

CT is often the first-line imaging modality for most soft tissue masses and EES. At non-enhanced CT, the solid components will typically have a similar attenuation to that of muscle. With contrast-enhanced CT, larger tumors will usually

show heterogeneous enhancement, with the hypoenhancing regions corresponding to areas of necrosis. Faint amorphous calcification occurs occasionally (25%-30%) and is more evident at CT than at other modalities. At MRI, EES tumors usually contain areas of internal high T2-weighted signal intensity corresponding to necrosis or cystic change. Chest CT and fluorine 18 FDG PET/CT are most sensitive for detecting lung and other distant or nodal metastases. PET/CT can be a useful adjunctive or even the main modality, especially after the diagnosis of EES is made. With fluorine 18 FDG, most sites of tumor involvement will show increased radiotracer uptake. This can especially be useful for staging and the detection of nodal and distant metastatic disease, which may alter management. Although bone scintigraphy has historically been used to help detect metastases in the skeleton, it has largely been replaced by PET/CT, which has a higher sensitivity for osseous macrometastases [2].

Treatment & Prognosis

The biopsy should be preferably taken from the extraosseous component to prevent pathologic fracture [8]. Since EES frequently contains areas of necrosis or hemorrhage, imaging can be used to target the most metabolically active portion for percutaneous biopsy, which is reflected by either tumoral contrast enhancement or fluorodeoxyglucose (FDG) avidity [2]. So biopsy determines the diagnosis. The biopsy cannot indicate the origin of the disease, as in our patient. At least an 18-gauge needle is preferred in percutaneous biopsy, with a significantly increased diagnostic yield with longer biopsy specimens, at least up to 10 mm. A minimum of three cores is desired to provide sufficient tissue for immunohistochemical and genetic testing [10]. The vast majority are heterogeneous (Figure 4), reflecting tumor necrosis or hemorrhage, although smaller tumors may appear more uniform. A large tumor with central necrosis that does not cross the midline is typical. Tumor cells demonstrate strong membranous positivity for CD99 (a cell surface glycoprotein) (Figure 4), which is almost universally expressed by ESFT cells at high levels but is not specific to ESFT [1]. The most common immunohistochemistry findings are staining for CD99 and no staining for TTF-1, cytokeratin, desmin and S-100 [1]. EES arises outside of the bone, and there should be no osseous component [1]. The absence of bone tissue in the pathological sample taken from the left lung mass of our patient led to the initial interpretation that it was PES (Figure 4). ESFT is often periodic acid–Schiff (PAS) stain positive and diastase sensitive, which can help differentiate it from other tumors such as lymphoma. In addition, immunohistochemical staining for FL-1, ERG, NKX2.2, and vimentin (less frequently) is usually positive [2].

Before chemotherapy, 10% of patients survived; however, 5-year survival for patients with localized disease now exceeds 70-80% with appropriate chemotherapy. Unfortunately, there has not been commensurate improvement in the survival of patients with metastatic disease, and their outcomes remain poor, with only modest improvement in 5-year survival ranging from 15-32% [2]. The disease often locally advanced, treated generally with multidisciplinary treatment combining surgery, chemotherapy and radiation therapy. At 36 months after diagnosis, only 16.7% of patients survive. 8.3% of them have been recovered of the disease in 48 months [1]. Determining the primary origin with advanced imaging, like our patient, will increase survival times.

Approximately one-fourth of patients with ES will have detectable metastases at presentation. As in ESB, EES metastases are most common to the lungs followed by bone or bone marrow [11-13]. First of all, the age of our patient, who was thought to have PES, was much younger than 9 years old, which is reported as the youngest age in the literature. Surgery was initially planned. Management often involves chemotherapy with local surgical excision, when possible [8]. Some cases with single resectable masses have also been described. Surgery, when feasible, is the best option for patients affected by PES/PNEt) [1]. Our patient is 3.5 years old and RT could not be performed due to the location and extent of the tumor and its location in the vital soft tissue structures (heart) region. Next is local control with surgery as the preferred mechanism, as it confers improved survival over that of radiation therapy alone. A multidisciplinary treatment approach should be used given the propensity for large tumor size and local invasion, which can make resection difficult [2]. Once diagnosis has been confirmed and staging completed, management is usually sought with local control (surgery and/or radiation therapy) plus chemotherapy with comparable outcomes between ESB and EES, including no difference in overall survival [8]. Surgical resection with wide margins yields a better 5-year event-free survival than with inadequate margins, although this benefit has not been consistently demonstrated in extremity tumors. Determining factors for resection feasibility include tumor location and extent, involvement of vital soft-tissue structures or the neurovascular bundle, the ability to completely excise the tumor with acceptable margins, and feasible reconstruction possibilities [2]. For those tumors felt to be resectable with negative margins and acceptable long-term physical function, definitive surgical local control is generally preferred to radiation. Definitive surgical resection typically takes place after several cycles of chemotherapy [8]. Unfortunately, upfront surgery can be performed in a limited number of cases. The treatment method was applied to only 14.7% of the reported cases [1]. Tumors that initially appear unresectable may become resectable with chemotherapy [8]. In our case, surgery was abandoned and chemotherapy was started due to metastatic nodules in the lower and upper lobes of the right lung after PET. In this situation, surgery would be meaningless and would be harmful as it would leave the patient without treatment during the recovery period. Metastases to the liver and brain are infrequent. Distant metastases are rare, as the disease tends to grow rapidly and to invade intrathoracic organs and chest wall anatomical structures. Although previously considered rare, regional lymph node involvement has been increasingly reported and may represent differing nodal involvement patterns for EES versus ESB. Lymph nodes were reported as the most common site of metastases in a recent large study of 70 EESs in an adult

population [1,11-13]. Due to the mass shrinking after the first course of ICA, it was understood that the main site of ES was the rib and that it had metastasized to the lung. Surgery could not be performed due to metastasis. The surgery planned to be performed; it was postponed to a later date after chemotherapy. Unfortunately, conventional cytotoxic chemotherapy is ineffective in up to one-fourth of patients with localized tumors and three-fourths of patients with metastases. In patients with metastases, attempts to add other chemotherapeutic agents or increase the dose have resulted in increasing toxic effects, without significant improvement in disease control or survival. Due to our patient's young age, in terms of secondary malignancies; A more controlled treatment plan was drawn. Megatherapy (myeloablative high-dose chemotherapy followed by stem cell rescue) has been attempted in patients with metastases outside of the lungs and pleura with mixed results. Following the standard therapeutic regimens, there is a small risk of secondary malignancy, with 1-2% of survivors developing acute myeloid leukemia or myelodysplastic syndrome [14]. A robust initial positive response to neoadjuvant therapy or irradiation predicts a better outcome and is associated with a decrease in the size of the primary mass. Decreasing tumoral contrast enhancement and increasing areas of necrosis or hemorrhage at MRI suggests a favorable treatment response. The definition of a positive response is greater than 90% tumoral necrosis at pathologic analysis, but unfortunately imaging is not sensitive to this degree. FDG PET avidity is regarded as a reliable marker of disease activity, with favorable treatment response reflected by decreasing tracer uptake. Persistent bulky tumors with decreased metabolic activity remain indicative of a positive response and correlate well with good histologic response, as well as improved survival rates [2, 12].

In most cases, a multidisciplinary treatment approach is chosen, integrating surgery, chemotherapy and radiotherapy in various combinations (61.2%). But less than 10% overcome 48 years of survival without signs of disease recurrence [1]. Clinically apparent metastases (macrometastases) as well as submicroscopic tumor in blood or marrow (micrometastases) are both indicators of a poorer prognosis. The presence of multiple metastases also portends a worse prognosis than the presence of a single metastasis. Patients with isolated lung or pleural metastases fare somewhat better than those with isolated bone or bone marrow metastases, who in turn fare better than those with combined lung and bone and/or marrow metastases, with cure rates of approximately 32%, 23%, and less than 15%, respectively [15]. Our patient had intact chest wall soft tissues and isolated lung metastasis; may result in a better prognosis. Neoadjuvant chemotherapy İs initiated with the hope of eliminating micrometastases and reducing the size of the primary tumor [8]. Neoadjuvant chemotherapy is employed in 30.6% of cases to downstage the disease and allow surgical removal. Adjuvant chemotherapy and radiotherapy have been employed with a certain success in some cases. In 12.2% unresectable cases, combination of chemotherapy with radiation therapy has been used. Unfortunately poor results have been obtained [1].

Although ESFT is considered radiation sensitive, the use of radiation therapy has declined owing to complications, which are more prominent in skeletally immature patients. Currently, radiation therapy is preferentially used to promote local control in primary lesions that are not resectable, in cases with inadequate surgical margins, or in patients with poor response to chemotherapy [16]. Due to our patient's young age, RT would increase all these complications. The decision to use radiation should be made with the understanding of the risk of secondary radiation-induced malignancies and the possibility of slowing of skeletal development and deformity [8]. Conversely, pulmonary metastases are optimally treated with whole lung radiation therapy rather than surgical resection of metastases [11]. Radiation is also used for metastatic disease and palliation of recurrent disease. Whole-lung radiation therapy with a dose of 12-21 Gy for patients with pulmonary metastatic EFT has been recommended as standard. Current recommendations for definitive radiation of the primary site utilize doses ranging from 45 to 60 Gy. In the postoperative setting, gross disease standard dosing is 55.8 Gy and microscopic disease is 45 Gy [8].

Differential Diagnoses

Future techniques incorporating biologic agents that target tumorigenesis, growth, or the development of metastases could be used in synergy with standard therapeutic regimens. Molecular therapeutics directed at cytogenetic tumor aberrations are varied in their approach and include (a) inhibition of the fusion gene or protein product, (b) inhibition of pathways or mediators in tumorigenesis (eg, tyrosine kinases, p53 pathways, CD99 expression), and (c) interference with angiogenesis and exploitation of nonapoptotic cell death. The multipronged approach has the potential to yield therapeutic regimens that are more effective, more selective, and less toxic in patients with ESFT [2, 17].

CONCLUSION

Determining the primary focus with appropriate imaging ensures correct treatment. Rare cases diagnosed with insufficient follow-up time and without advanced imaging methods should be questioned again.

TEACHING POINT

Determining the primary focus with appropriate imaging ensures correct treatment. Rare cases diagnosed with insufficient follow-up time and without advanced imaging methods should be questioned again.

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FIGURES

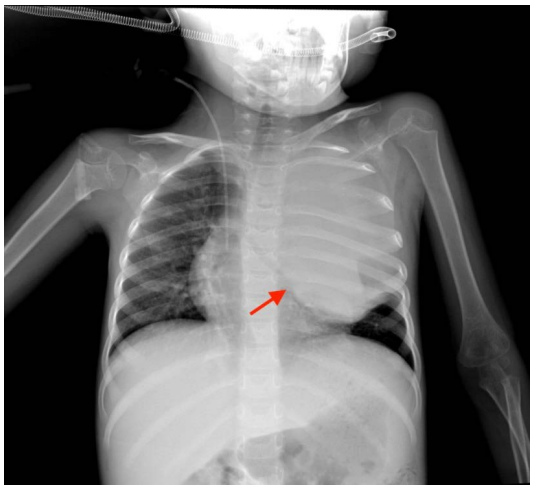

Figure 1: Radio-opaque mass in the upper area of the left lung in direct radiography

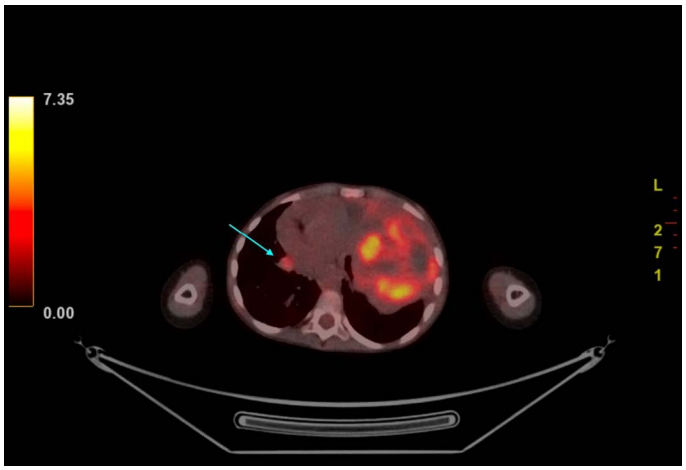

Figure 2: PET/CT are most sensitive for detecting lung and other distant or nodal metastases

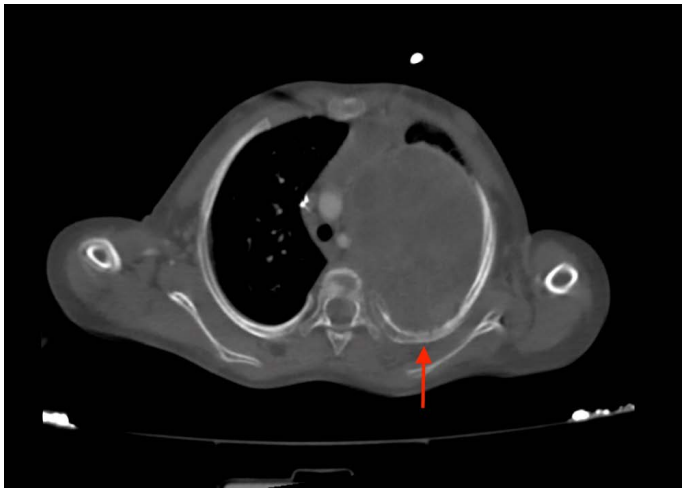

Figure 3: Diagnosis of PES with CT after the first ICA; It changed to ES, which metastasized from the ribs to the lung (solid lesion originating from the posterior of the left 3rd rib, with intact mediastinum and lung intermediate planes, 71X93X89 mm with well-defined borders and necrotic areas).

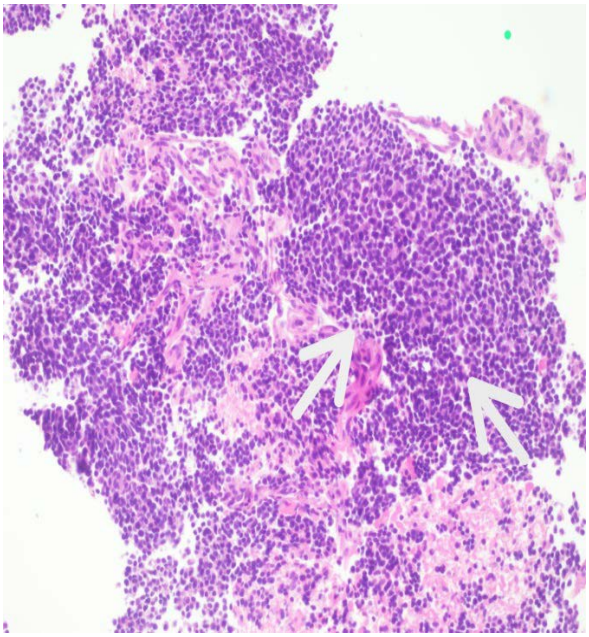

Figure 4A: x20 magnification Hematoxylin-Eosin

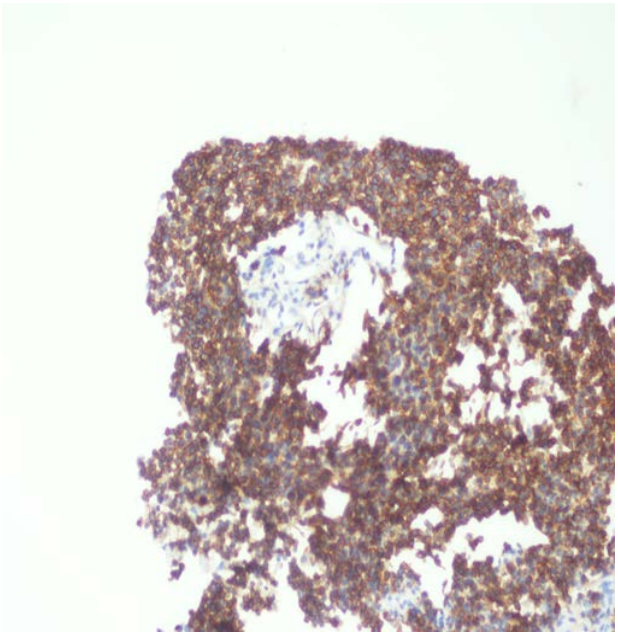

Figure 4B: x20 magnification CD99

Figure 4: Tumor cells showing diffuse membranous positivity for strong CD99 staining (no staining for TTF-1, cytokeratin, desmin and S-100)

KEYWORDS

Ewing Sarcoma, Extra-Skeletal Ewing Sarcoma, Thoracopulmonary Ewing Sarcoma, Primary Pulmonary Ewing Sarcoma

ABBREVIATIONS

ES = Ewing Sarcoma
EES = Extraskeletal ES
PES = Primary pulmonary ES
PET = Positron emission tomography
FDG = fluorodeoxyglucose
VDC = Vincristine, doxorubicin, and cyclophosphamide
ESFT = ES family of tumors
EWSR1 = Aberrant fusion of the ES gene