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Extraskeletal Ewing's Sarcoma of the Infratemporal Fossa with Orbital and Intracranial Extension: A Rare Tumor in A Rare Location: A Case Report

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ABSTRACT

Extraskeletal Ewing's Sarcoma is a rare soft tissue tumour morphologically similar to the commoner Ewing's Sarcoma arising from bone. Head and neck origin of this tumour is rarely observed with infratemporal fossa Ewing's Sarcoma forms another uncommon subset. We report a case of Extraskeletal Ewing's Sarcoma in the infratemporal fosssa in a 5-year-old girl; who presented with a rapidly growing mass in the left cheek. CT scan of neck and head showed soft tissue mass in left infratemporal fossa with intraorbital and intracranial extention. Histopathological examination of the biopsy specimen showed round cell tumour and immunohistochemistry was positive for CD99 and Vimentin. According to the clinical manifestation, CT findings, histological pattern and the results of the immunohistochemical studies, the final diagnosis was Extraskeletal Ewing's Sarcoma. Our patient was initially treated with chemotherapy. The patient responded very well to chemotherapy. For the management suite, a radiotherapy is programmed. To our knowledge, this is the second case of EES in infratemporal fossa in paediatric age group. Early and certain diagnosis coupled with combined surgical excision and modern chemotherapy/radiotherapy seems to be the most effective treatment plan.

Keywords: Extraskeletal Ewing's Sarcoma; Infratemporal fossa; Child; CT scan; Immunohistochemistry

INTRODUCTION

Ewing's Sarcoma (ES) is a malignant small round blue cell tumor that develops from primitive neuroectodermal cells and occurs primarily in bone or soft tissue. It was first described in 1921 by James R. Ewing.[1] The ES

family of tumors now includes ES of bone, extraskeletal ES, Askin tumors of the chest wall, and primitive neuroectodermal tumors (PNET) of bone or soft tissues.[2] Extraosseous sarcomas account for about 15% of all Ewing's sarcomas.[3] The paravertebral region was the most frequently involved region, followed by the lower extremity and chest wall. The head and neck origin of this tumor is uncommon, accounting for only 1-4% of all cases of Ewing's Sarcoma,[4] with infratemporal fossa Ewing's Sarcoma forms another uncommon subset. It has histological, immunohistochemical, and molecular characteristics that are similar to bone Ewing's Sarcoma. According to most authors, ES of the head and neck has a better prognosis than those that arise in other locations.[4] This article describes the clinical, radiological, histopathological, and immunohistochemical features of a rare case of Extraskeletal Ewing's Sarcoma (EES) originating from the infratemporal fossa with intraorbital and intracranial extention. An extensive literature search found only one case of Extraskeletal Ewing's Sarcoma in the infratemporal fossa in a 13-year-old female reported by Lee, R.J et al.[5] We present the second case of infratemporal fossa Extraskeletal Ewing's Sarcoma.

CASE REPORT

A 5-year-old girl with no known medical or chirurgical history, presented with a rapidly growing mass in the left cheek since past 3 months. Clinical examination found a mass which was rubbery, mild tenderness, and fixation to underlying structures. The overlying skin was of normal texture and color. No palpable cervical lymphadenopathy was present or hepatosplenomegaly. The cranial nerves were intact.

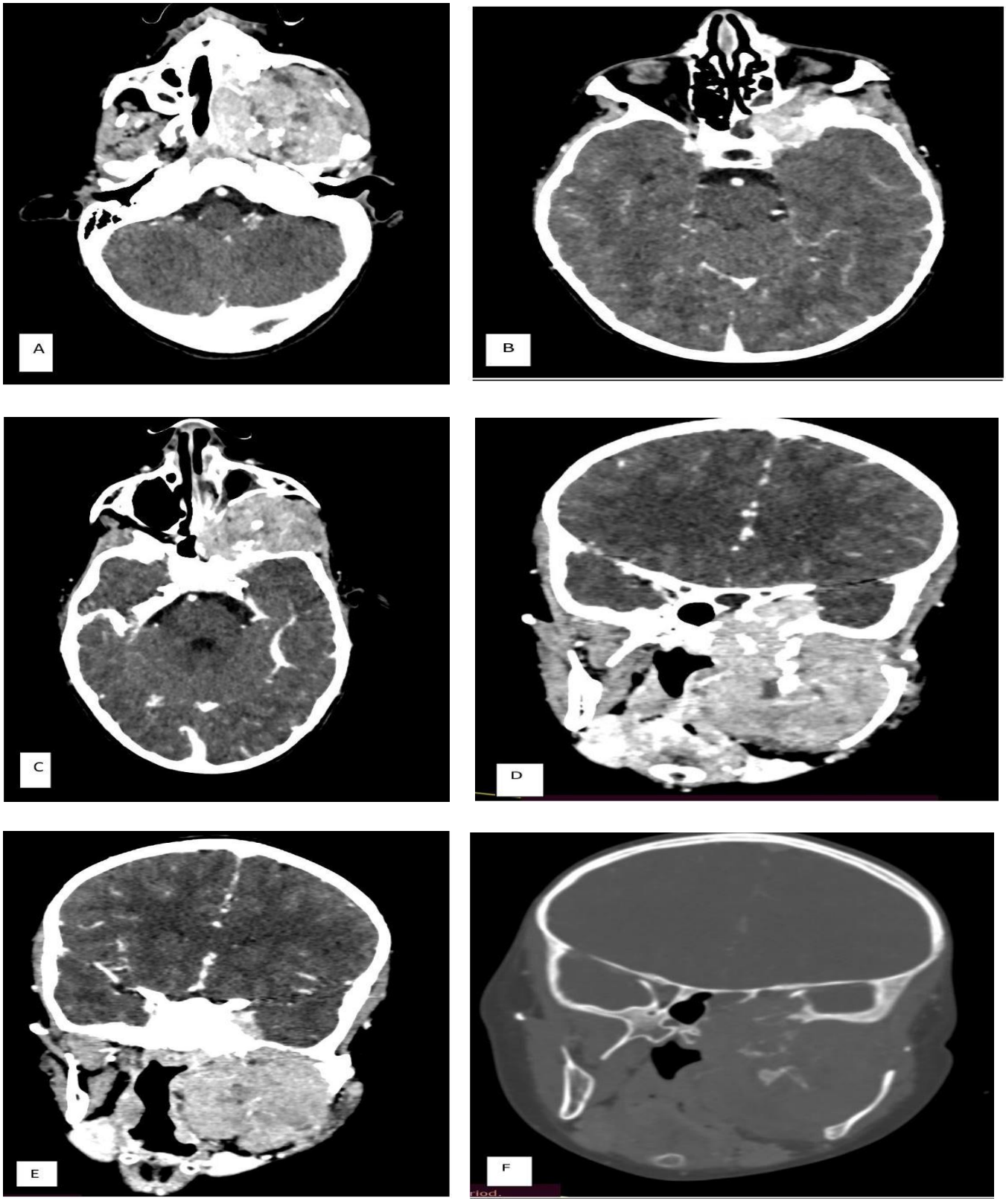
Computed tomography (CT) neck and head with contrast demonstrated a poorly enhancing expansile mass in the left infratemporal fossa region (ITF) with areas of necrosis within. The tumor was seen to extend to the left sphenoid sinus, left ethmoid sinus, nasal cavity, left parapharyngeal space, left pterygopalatine fossa and left orbital cavity. There was also destruction of skull base and extension to the left foramen ovale, left cavernous sinus and left temporal lobe. The mass appeared to have caused some bony erosion of the posterior aspect of the left maxilla. The left maxillary antrum was reduced in volume. It thins the left mandibular ramus and the left zygomatic process without cortical lysis and extends to the soft tissue of the cheek. The lateral pterygoid process was expanded, the median pterygoid process was eroded. No obvious calcification or aggressive periosteal reaction seen. Radiological diagnosis was malignant tumor of the infratemporal fossa. (Figure 1)

She subsequently underwent a biopsy of the mass. Histopathological examination showed features consistent with small round cell tumour. The tumour demonstrated high cellularity composed of small, round uniform cells with oval nuclei and scanty cytoplasm (Figure 2). Some of these cells are forming rosettes. The cytoplasmic organelles were poorly developed. Panels of immunohistochemical tests were done showing the neoplastic cells were positive for CD99 (Figure 3) and Vimentin (Figure 4), and nonreactivity to Cytokeratin, actin, neuron specific enolase and leucocyte common antigen. Thus histological and immunohistochemical findings were confirmatory for Ewing's Sarcoma.

She was then referred to department of pediatric oncology for further investigation and proper treatment. The studies to exclude metastatic disease (CT scan of thorax and abdomen, skeletal radiography study, bone studies with radionucleotide, bone marrow biopsy) were negative.

In view of the extensive tumour, the patient was treated according to `EURO EWING 99` protocol with induction chemotherapy consisting of vincristine, ifosfamide, doxorubicin and etoposide planned for 6 cycles,

21 days in between. Later, a follow-up CT scan showed significant reduction of the tumour as well as improvement in local invasion after 6 cycles of chemotherapy (Figure 5). For the management suite, a radiotherapy is programmed.



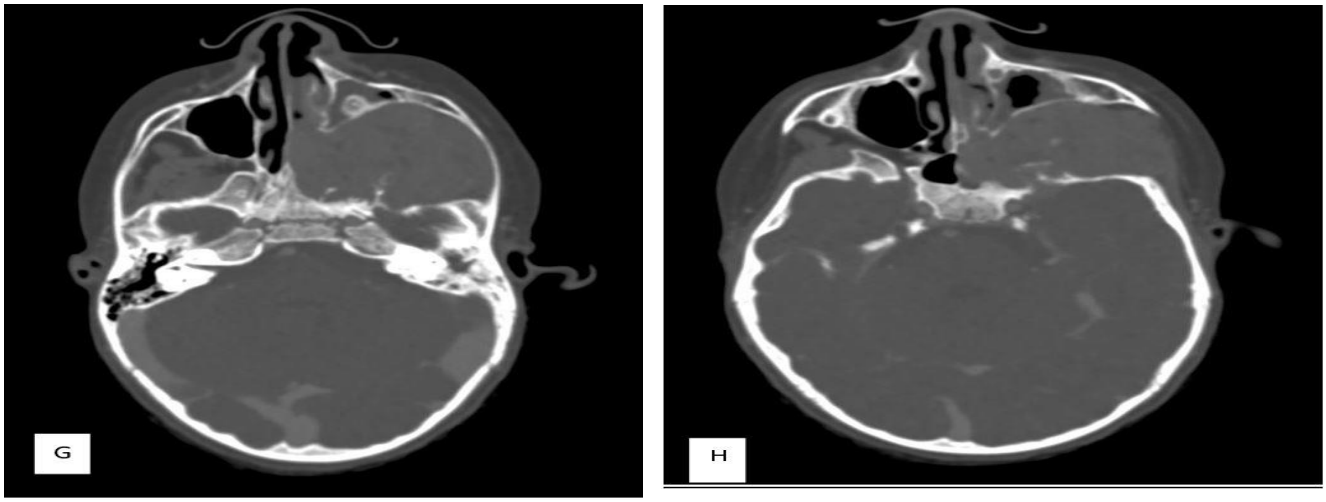


Figure 1: : (A-E) Axial and oronal and cut of contrast-enhanced CT scan of the neck and head, and (F-H) bone-reconstruction CT. Computed tomography (CT) neck and head with contrast demonstrated a poorly enhancing expansile mass in the left infratemporal fossa region(ITF) with areas of necrosis within. The tumor was seen to extend to the left sphenoid sinus (Fig.D)., left ethmoid sinus, nasal cavity (Fig.A)., left parapharyngeal space, left pterygopalatine fossa (Fig.A). and left orbital cavity (Fig.B). There was also destruction of skull base (Fig.G) and extension to the left foramen ovale (Fig.A)., left cavernous sinus and left temporal lobe (Fig.E). The mass appeared to have caused some bony erosion of the posterior aspect of the left maxilla (Fig.H). The left maxillary antrum was reduced in volume (Fig.C). It thins the left mandibular ramus and the left zygomatic process (Fig.F).without cortical lysis and extends to the soft tissue of the cheek. The lateral pterygoid process was expanded, the median pterygoid process was eroded (Fig.F).

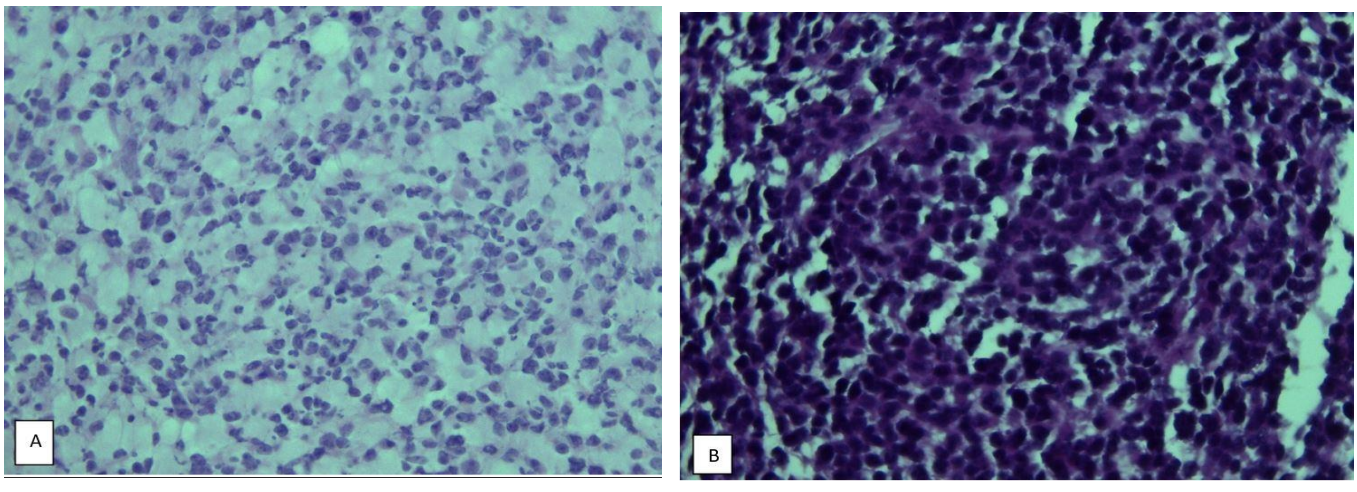


Figure 2: The infratemporal fossa mass is composed of monomorphic small roundblue cells arranged in diffuse sheets displaying monomorphic , round hyperchromatic nuclei, absence of nucleoli. Some of these cells are forming rosettes (A : :Hematoxylin and eosin, 200x magnification, B :Hematoxylin and eosin, 40x magnification)

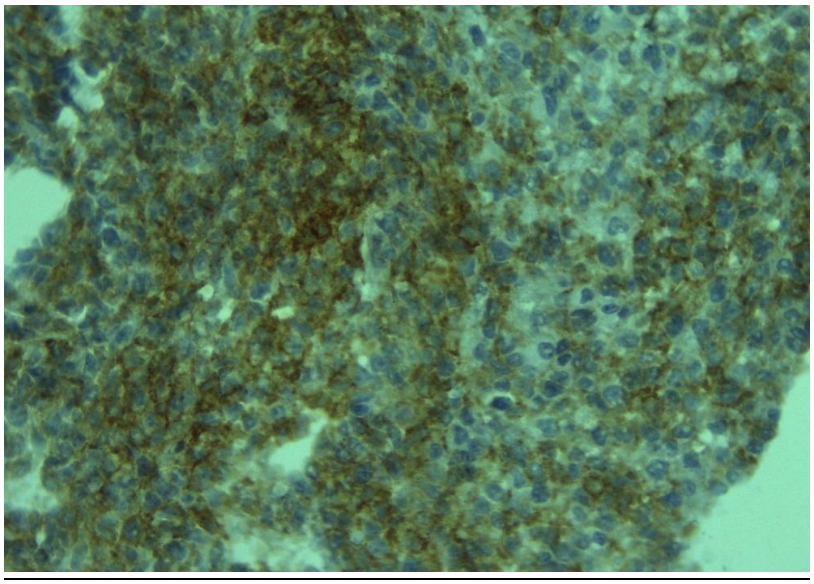


Figure 3: The malignant cells are diffusely positive for CD99 (40x magnification)

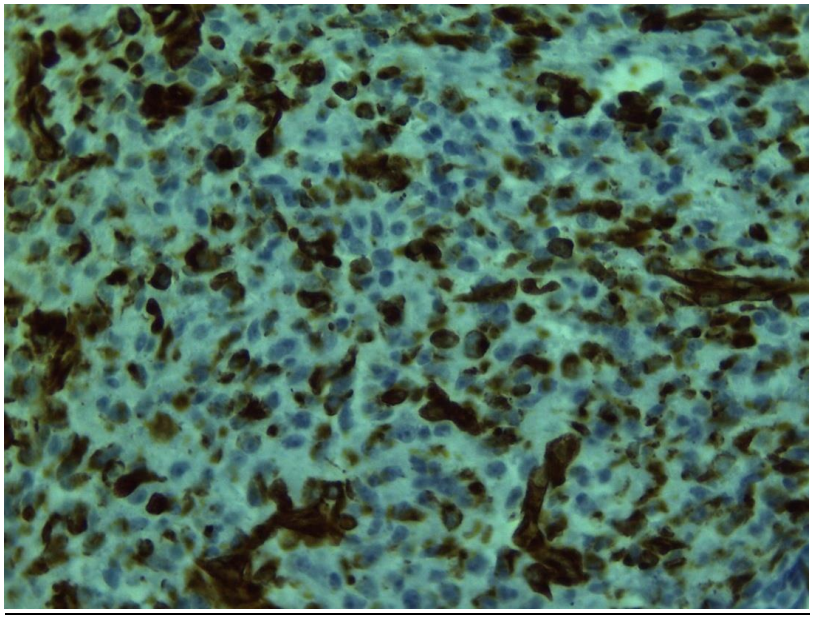
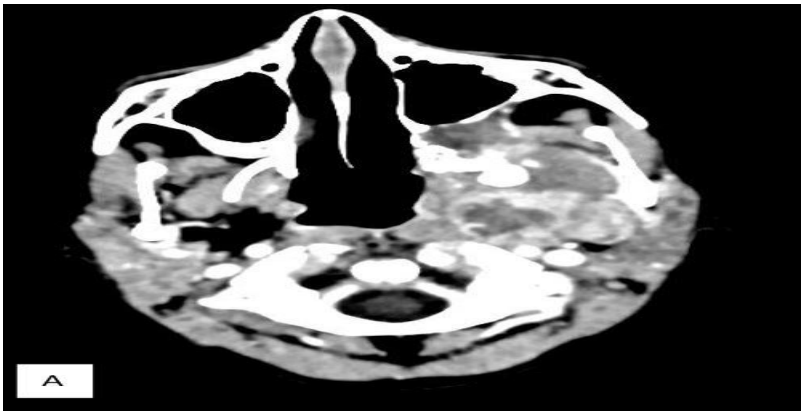


Figure 4: The malignant cells are diffusely positive for Vimentin (40x magnification)



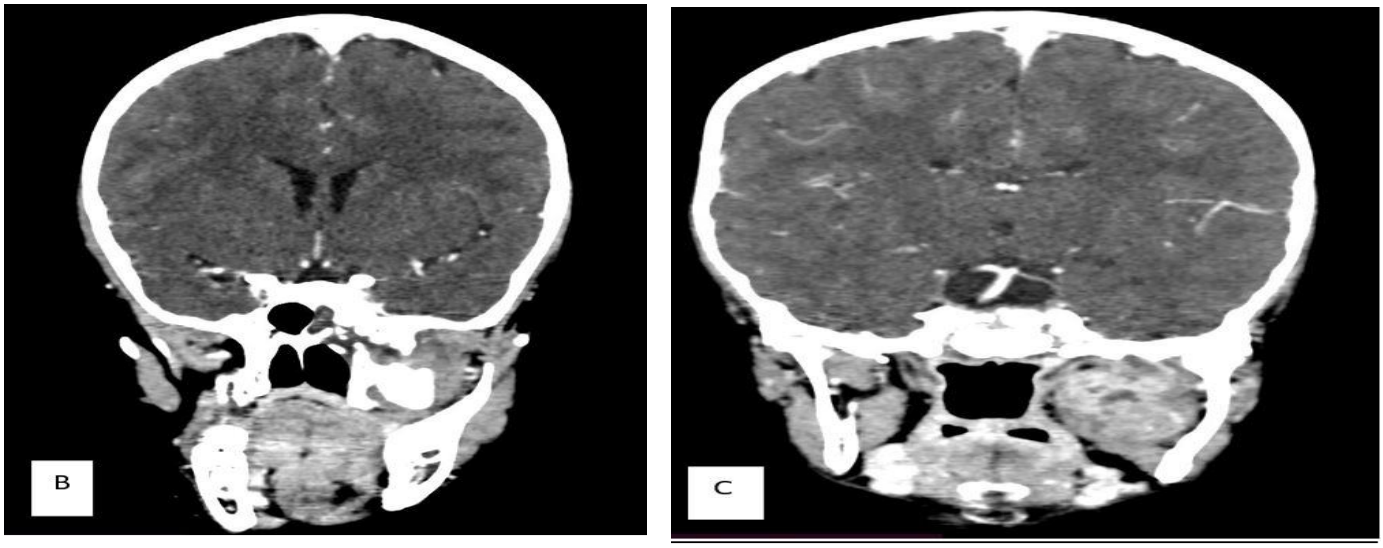


Figure 5 : Axial (A) and coronal (B, C)cut of contrast-enhanced CT scan of the neck and head after 6 cycle of chemotherapy, shows significant reduction of tumour size and its mass effect. The tumour in (5) shows more peripheral enhancement with poor enhancement centrally in comparison with prechemotherapy in (1).

DISCUSSION

Extraskeletal Ewing sarcoma (EES) is a rare tumor that belongs to the ES family of tumors (ESFT), which is a group of small round tumor cells with a common neural histology and genetic mechanism.[6] Tefft et al. first described EES in 1969.[7] It primarily affects adolescents and young adults aged 10 to 30 years, but cases of patients aged between 14 months to 63 years have been reported.[4] There is no sex predilection.[7] Although it can develop in any soft tissue, the head and neck origin of this tumor is extremely rare, accounting for only 1% to 9% (8), with cases reported in the orbit, scalp, face, nasal cavity, paranasal sinus, nasopharynx, parapharyngeal space, larynx, hard palate, submandibular gland, parotid gland, thyroid gland, pterygomandibular space, and soft tissue of the neck.[3] Only one case has been reported in the infratemporal fossa.[5]

There is no specific clinical manifestation of EES in the head and neck region. The most common symptom is a rapidly growing painless mass. However, the type of accompanying symptoms depends largely on the site of involvement of the tumor and may include dysphagia, hoarseness, dyspnea, and limited range of mandibular motion. At the time of diagnosis, about 30% of patients had distant metastasis.[4,9,10,11] Our patient presented with a rapidly growing painless mass in the left cheek but no distant metastasis.

Imaging is extremely crucial in the diagnosis, staging, treatment monitoring, and surveillance of EES.[12] However, the imaging features of EES are non-specific.[2,12,13] EES is typically appears on ultrasound as a heterogeneous mass with low echogenicity and intratumor flow signals on a Doppler study.[2] EES appears on CT as a large sharply demarcated mass with the same intensity as the surrounding muscles. The enhancement of the post-contrast medium reveals areas of necrosis, while the surrounding viable contour appears enhanced and heterogeneous. Only 10% of cases have calcification, which appears faint and amorphous. EES has a low-to-intermediate signal intensity on T1-weighted sequences and a high signal intensity on T2-weighted images, with

variable post-contrast enhancement on MR imaging. Prior to biopsy, MR imaging is used to help determine the best biopsy site and to prevent distortion caused by post-biopsy changes. This is also recommended prior to local control because the tumor may have receded or progressed during neoadjuvant chemotherapy.[2] In addition to local tumor staging, imaging is used to detect the presence of metastatic disease. Chest CT scans to search for lung metastasis. FDG-PET is used to detect bone metastasis as well as to assess chemotherapeutic response and detect recurrent disease.[2] The lung is the most common site of metastatic disease in patients, followed by bones and bone marrow.[9]

A histopathological examination, immunohistochemistry, and cytogenetics are required for a definitive diagnosis.[7,12] EES is histologically similar to osseous ES and is frequently confused with other round cell tumors, but immunohistochemistry advances have aided in the diagnosis of ESS. EES appears as monomorphic, small, round blue cells with large spherical nuclei, inconspicuous nucleoli, and indistinct cytoplasmic borders under the microscope.[2] In ESFT, there is a strong immunoreactivity for CD99 and vimentin. Negative staining for Leucocyte common antigen, CD30, actin, Neuron specific enolase, and S-100 enables us to exclude the diagnosis of lymphoma and rhabdomyosarcoma. The absence of neurosecretory granules (neurofilaments) distinguishes classic ES from primitive neuroectodermal tumors PNET. Pathology and immunohistochemistry are usually sufficient to diagnose EES. In the case of unusual variants, molecular genetic analysis using reverse transcriptase (RT) PCR or fluorescence in situ hybridization (FISH) can be used. The two most common EES and ESFT chromosomal translocations are t(11;22)(q24;q12) and t(21;22) (q22;q12). (2), which are seen in 85-90% of ESFT cases.[4] In our patient immunohistochemistry revealed round cells with high cellularity with scanty cytoplasm and positive reactivity for CD99 and vimentin.

Our case had lymphoma and rhabdomyosarcoma as differential diagnoses, which were all excluded on by radiological, histological, and immunohistochemical findings. This unusual case of infratemporal fossa EES demonstrated that immunohistochemistry is a very useful aid in the diagnosis of aggressive deep-seated tumors of the head and neck region, which aid in early diagnosis and timely intervention, resulting in reduced morbidity and mortality.

Anderton et al. proposed the Euro Ewing 2012 protocol for treatment of ES based on age at diagnosis, disease stage (localized tumor, presence of lymph nodes, or distant metastasis), and tumor volume at diagnosis.[14] With current multimodal therapy, which includes combination chemotherapy, surgery, and radiotherapy, a localized ES has a 5-year survival rate of about 65%, and a 3-year event-free survival (EFS) rate of 30% in patients with ES and lung-only metastases. Patients with disseminated disease had an extremely poor prognosis for this tumor, with only 29% achieving 3-year overall survival (OS).[14]

CONCLUSION

Although ES of infratemporal fossa is very uncommon, it may be considered in the differential diagosis of infratemporal fossa tumors with intracranial and intraorbital extension in adolescents and young adults. its diagnostics in this localization is challenging. CT and MRI cannot provide a specific diagnosis, but successfully demonstrate the internal structure of the lesion and the extent of the tumor. Early and confident diagnosis coupled with combined surgical excision of gross disease and modern chemotherapy/radiotherapy appears to be the most effective treatment plan.

AUTHOR'S CONTRIBUTIONS

All authors contributed to this work. All authors have read and approved the final version of the manuscript.

CONFLICTS OF INTEREST

The authors declare that there are no conflicts of interest regarding the publication of this paper.

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