ISSN: 2581-5989
PubMed - National Library of Medicine - ID: 101738774

International Journal of Dental Science and Innovative Research (IJDSIR)

IJDSIR : Dental Publication Service

Available Online at: www.ijdsir.com

Volume – 5, Issue – 6, November - 2022, Page No. : 208 - 214

Ewing's Sarcoma of Infraorbital - A Case Report

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Citation of this Article: Ahmad M. Altarawneh, Amjad M. AlWarawreh, Reham A. Hijazen, Mohammad S. AlQaralleh, Yasmin M. AlSaidat, "Ewing's Sarcoma of Infraorbital - A Case Report", IJDSIR- November - 2022, Vol. – 5, Issue - 6, P. No. 208– 214.

Type of Publication: Case Report

Conflicts of Interest: Nil

Abstract

Background: There are a large variety of non-epithelial tumors, and Ewing sarcoma is one of them. It is neural or neuro-ectodermal in origin. It is a malignant tumor that is composed of small round cells of bone and soft tissues. It is very aggressive in nature (1) and is considered to be 6th most common among deadly tumors. The aggressiveness of the tumor is on account of its undifferentiated cells. 10 ton15% of all the primary malignant tumors are Ewing sarcoma and it is also the 2nd leading malignant tumor which was found to be common in children and adults (2).

Neuroblastoma, rhabdomyosarcoma, desmoplastic small round cell tumor, lymphoma and the Ewing family of tumors (ESFT), all belong to category of tumors of small round cells in the childhood (3). They all are highly aggressive in their growth and have potential of malignancy and when observed under the microscope, they appear as a homogenous growth of small cells that have a scanty cytoplasm (4). James Ewing in 1920 described the Ewing sarcoma as a diffuse growth of endothelial bone cells (5). The undifferentiated mesenchymal cells of the bone are considered the primary origin of Ewing sarcoma in adolescents as well as young adults, ranging in age from 10 to 20 years (6,7). The incidence of Ewing sarcoma before the age of 50 and after the age of 30 years is considered as a rarity. There is a slight male preponderance with the male to female ratio of 1.5:1 (1). The genetics also play a role as a sibling of a patient suffering from Ewing sarcoma has

more likelihood to develop one too. The origin of Ewing sarcoma has always been a controversy as some believes it comes from endothelial bone cells, while others consider it of primary mesenchymal origin of bone marrow. The other theories believe that it comes from immature reticular cells while some thinks of it as of neural origin(8).

The Ewing sarcoma targets the skeletal system and it makes up 4 to 10% of all the long bone cancers(2), femur being the most commonly affected bone. However, it rarely occurs in head and neck as its prevalence is less than 3% in these regions(9), however mandible and then maxilla being the commonly affected bone in maxillofacial regions. The site of the tumor, the size od the tumor mass and the status of metastasis are the major prognostic factors (10). With the progress of the disease, cortex of the bone gets destroyed and then the mass invades the periosteum and enters the local soft tissues(11).

A case report will be presented to get the in-depth analysis of the clinical picture as well as radiological landmarks (CT) of a case of Ewing sarcoma along with the histopathological biopsy results of a 14-year-old female who developed Ewing's sarcoma of the infraorbital wall.

Case Description

An 14-year-old female child presented to the Dental Department of Prince Ali ben AL Hussein Hospital in Karak City in Jordan in Aug 2021, with a rapidly enlarging swelling on right infra orbital region since one month. There was no history of trauma or pain associated with the swelling. Also, no secondary or sensory changes could be elucidated. her past dental/medical history was unremarkable.

The extraoral examination a sudden onset swelling in right infra orbital side gradual increase in size over time. On palpation, the inspector findings were confirmed. Firm painless subcutaneous mass, limited mobility and slightly demarcated from surrounding tissue No skin adhesion, No loss of sensory input or motor function, Normal visual acuity of RT globe and Normal extra orbital muscles (EOM) motility of RT orbit.

The radiographic examination. CT sacn was advised so the site, extent and local effects of the tumor as well as any metastasis can be pinpointed. CT sections showed osteolytic expansile destructive bony lesion involving the inner aspect of the right anterior zygomatic bone lateral wall of right maxillary sinus and anterior orbital floor, associated with multilobulated heterogeneously enhancing soft issue component with necrotic no enhancing areas extending to the inferior aspect of the orbital cavity and right maxillary sinus(2.7 cmX2.6cm orbital soft tissue component,1.8x1.1 cm right maxillary component) both orbital globes and orbital rectus muscles and nerves are unremarkable bilaterally. Both lacrimal glands appear unremarkable. Fig 1

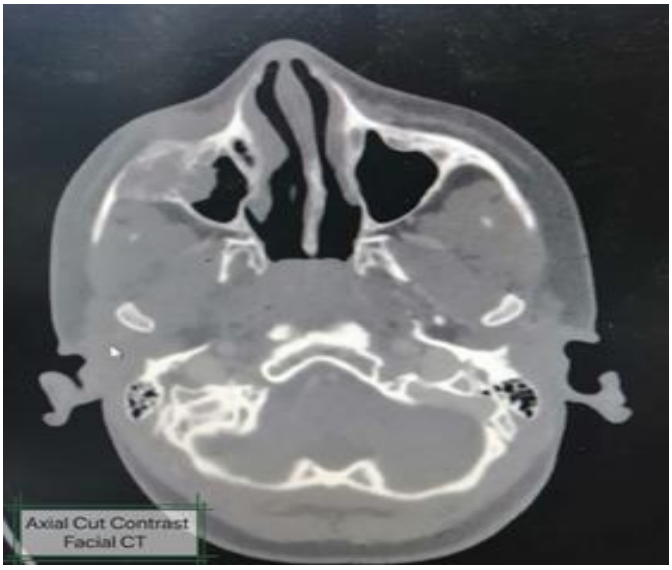

Fig 1: Axial cut of Brain CT with IV contrast, (Bone window). There is expansile lytic bone lesion with enhancing soft tissue mass arising from in ferio-lateral aspect of the right orbital wall, causing bone destruction of inferior orbital wall and right zygomatic bone

extending down to the right maxillary sinus. This lesion is associated with Codman triangles (type of periosteal reaction seen in aggressive bone lesions).

18F-FDG PET CT Whole body Scan was done and it was observed that there was no evidence of distant metastasis.

However, there was a mildly hyper-metabolic as well as destructive bony lesion at the site of right orbital bone inferiorly that invaded the associated soft tissue component. The mass extended to adjacent right maxillary sinus too. There was no other such lesion observed in the rest of the scan. (Figure 2)

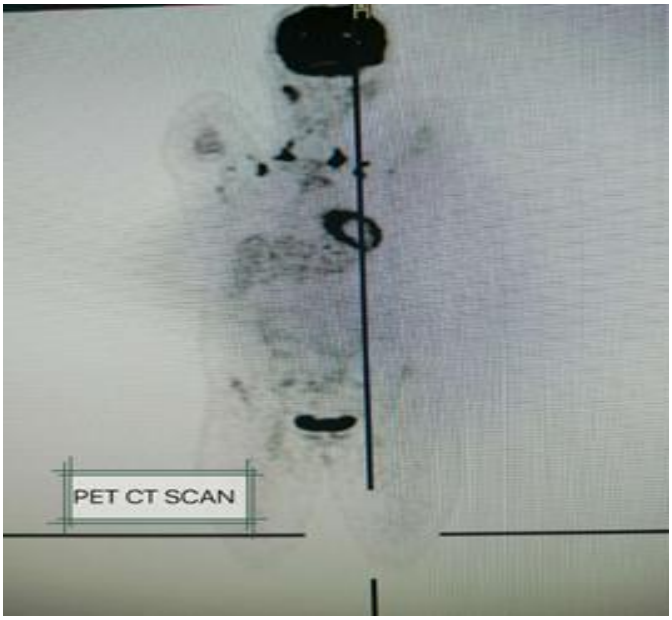

Fig 2: 18 F-FDG PET/CT whole body scan - Coronal cut.

Mildly hyper-metabolic destructive bone lesion seen in right orbital bone inferiorly with associated soft tissue component.

Orbital MRI with IV Contrast

When a T2-weighted image was studied in detail, there was an irregular shaped intermediate to high signal soft tissue mass that has ill-defined borders and looks like a malignant one. However, a hypointense intensity was observed on T1-weighted image that was present along with the heterogeneous enhancement that is usually bony in origin. This is located at the right anterior zygomatic bone, lateral wall of right maxillary sinus and orbital floor anteriorly and extend to right maxillary sinus and to inferior portion of orbital cavity that then gets compressed and deviates the orbital structures. However, there is no sign of local invasion observed. The mas was almost 2.7x3.4x2.3 cm in maximum dimension and there was no intercranial extension (fig 3)

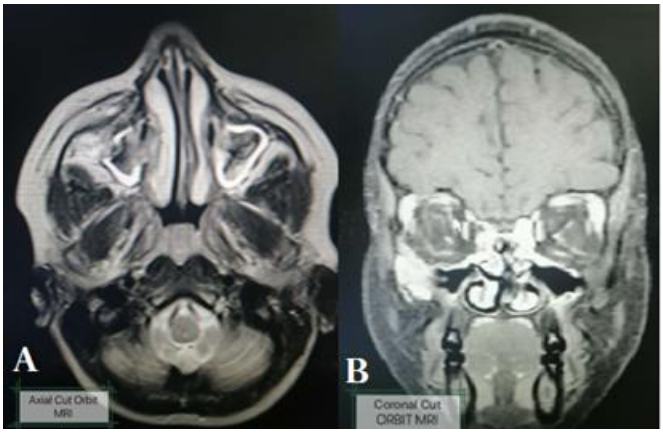

Fig 3: Axial cut (A) and Coronal cut (B) of T2-weighted image of Brain MRI showing hyper-intense lesion noted at in ferio-lateral aspect of the right orbital rectus muscle

Neck and cerebral CT Angio

An enhancing extraconal soft tissue lesion was seen in the inferio-lateral region of the right orbit that was facing the right inferior orbital rectus muscle and was about 3.4 in cm in AP dimension. 2.8 cm in transverse dimension and 2.5 cm in craniocaudal dimension and was leading to the mass effect. The effect cause pressure upon the right globe and cause it to displace superiorly and medially. This also lead to the destruction of the inferior orbital wall, right zygomatic and sphenoidal bone and even the right maxillary sinus. This confirms that the mass is rhabdomyosarcoma.

Incisional biopsy was also taken in the Feb, 2022. The diagnosis made was Ewing's sarcoma /PNET when histopathological tests was carried out. There were sheets of homogenous as well as small round cells that

were arranged in a diffuse pattern. There were no distinct borders, cytoplasm was scanty. Cells had a well-defined nuclear membrane and the cells were having nucleus of round to oval shape. However no mitotic figures were observed. (Fig. 4). Positive CD99 & Intracytoplasmic granules are positive for PAS and sensitive for PAS-D (Fig 5)

Fine needle aspiration: right and left bone marrow aspiration were taken show normal finding with no evidence of infiltrative marrow disease.

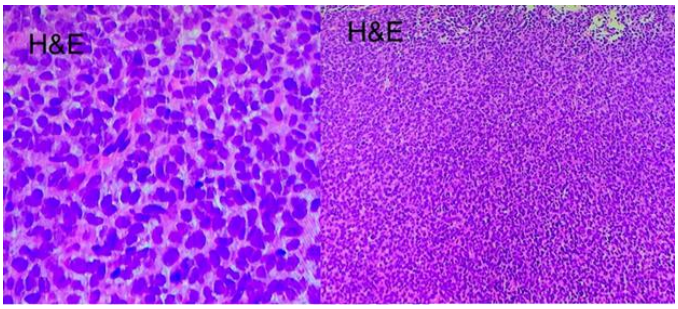

Fig 4: Hematoxylin and eosin slides reveal a highly cellular tumor composed of small monotonous round to oval cells with round nuclei and scant cytoplasm, the stroma of which is minimal and fibrovascular. Mitotic activity identified(3MF/HPF)

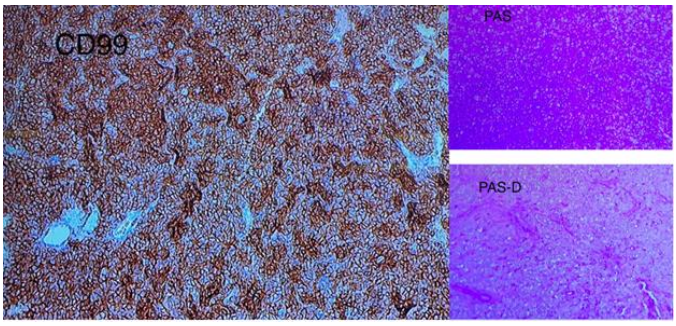

Fig5: CD99 immunostaining is strongly positive (membranous) in tumor cells. PAS stain highlights cytoplasmic glycogen in the tumor cells, diastase sensitive

Ancillary studies;

Myogenin, Desmin, S100, Tdt, synaptophysin, CK, CD56: Negative.

Fig 6: Obtaining a sample from tissues of abnormal growth is essential part of the diagnostic process for the purpose of his to-pathological examination. As shown above, patient underwent incisional biopsy procedure conducted under general Anesthesia, access to the tumor is achieved through sub-ciliary incision.

Fig 7: Surgical site closure performed by mean of primary intention using non-resorbable synthetic monofilament suture (nylon).

Fig 8: an operated site healing after 4 weeks from Surgery

Treatment

After incisional biopsy and confirmation of diagnosis. The standard treatment options were discussed with haematologist Pediatric oncologist according to protocol used in Royal Medical Services 1st line treatment

neoadjuvant chemotherapy for sake of tumor shrinkage and to facilitate surgery later on however based on tumor location extra coronal orbital floor without intracranial extension and as its abutting lateral rectus muscle ,surgical procedure were defer due to anticipated difficulty in reconstruction issues and possibility of visual impairment ,radiotherapy and chemotherapy and patient is on both treatment modalities

Patients were start chemotherapy on April 2022 and start radiotherapy in august 2022

After finishing both chemotherapy and radiotherapy the physician and patient feel improvement regarding the size and no visual disability present nor visual impairment report before and during treatment

Surgery were schedule for complete excision with safety margin but due to good response for chemotherapy and radiotherapy in addition to the complexity and risk of destruction of vital structure it was finally decide not to do any further surgery which indeed improve her quality of life.

Discussion

Ewing Sarcoma is a cancer of long bones of the limbs and half of the cases affect the femur and pelvis. The white have more chances of developing this. However, it is rare in blacks as well as in regions of head and nick is rare(9). A hem Angio-angiogenic origin has been suspected due to the structural similarity with the desmosomes and basement membrane or alkaline phosphatase activity. However, there was no evidence found that would support the angiogenic origin(11). However, studies by electron microscope and immuno-histochemical staining have suggested a neurogenic origin. Genetics has shown that there is a balanced translocation involving the EWS gene on chromosome 22 and a member of the ETS family of transcription factors(12). The disease usually starts in the Haversian canals, subperiosteally, or in the medulla of the long bones of the limbs. The clinical presentation is a mass that is highly locally destructive as well as expansible. The mottled radiolucency gives a laminated or onion skin periosteal reaction, is also observed and that is also the hallmark of ES. (13)

Histopathological there is glycogen stores in the cytoplasm that can be demonstrated by PAS staining. But it is not a specific finding as conclusive as presence of glycogen can be seen in other small round cell tumors also (9). Cytogenetic studies and molecular bio markings have made the diagnosis of Ewing sarcoma much easier and more accurate as well as rapid. To diagnose the overexpression of CD 99, a transmembrane protein that is encoded by the MIC-2 gene by immuno-histochemistry also play an adjunct role in this regard(6,12). According to Oberlin O, the embryonic reticulum cell that is present bone marrow are the precursor cancer cells as same dark cells are observed in a mature reticular portion of bone too (14). However, some other consider these dark cells to be a result of degenerative changes of the principal cell.

The clinical manifestation, morphology of the tumor, radiology, immuno cytochemistry and ultra-structural features of ES and ESFT play a key role in differentiating them from other small round cell tumors of childhood, such as neuro blastoma, rhabdo myosarcoma, Hodgkin's lymphoma, other primitive neuro ectodermal tumors, desmoplastic small round cell tumor, poorly differentiated synovial sarcoma, and small cell osteosarcoma(2).

The typical "histiocytic" features are the landmark of a eosinophilic granuloma and lymphoid cells along with the round cell of varying size are a feature of malignant lymphoma that have negative PAS staining and positive reticulum staining (15).

Treatment

Ewing sarcoma has well observed sensitivity to radiation therapy as compared to other malignant bone tumor (16,17,18) s. The combination of radiotherapy and chemotherapy first used by Rosen et al. in 1974 it resulted in improved long-term survival from 5% to10% to 50% to 60% during the past 30 years (18-23). Treatment of intra and extra cranial sarcomas are different as intracranial ones are aggressive locally and mental deficit demands urgent surgical intervention. However, resection cannot be done completely intracranially as the neoformation of them come across essential structures so a radical surgery is preferred. However radical surgery comes with affected quality of life and loss of function of the brain. Therefore, the level to which a resection can be done is determined by the site and size of the tumor mass. (16,17,20,24–25). Preparation of the tumor with the chemotherapy before the resection can lead to decrease in the site of the tumor and eliminates any micro metastasis.

If there tumor is inoperable, preoperative radiotherapy can be opted for. Radiotherapy is opted for patients who doesn't tolerate chemotherapy and the tumor excised is not enough.

However, there is no need of radiotherapy if patient is well tolerated with the chemotherapy and radical excision has been done adequately. Moreover, a primary ES give excellent response to radiation and chemotherapy, a radical surgical resection is also a good to go option (16,17,24,26–28).

The complications that occur late as well as secondary tumors are the major risks. Thus makes the long-term follow-up necessary in such patients.

Conclusion and clinically significant

A specific clinical, imaging, and histological feature accompanies the Ewing sarcoma. A through testing and a proper long-term follow-up is essential in such cases. Though a rare cancer, but it involves the facial bones of young individuals. Our case study highlighted clinical manifestation and various radiological features essential for the diagnosis of this rare tumor. If the swelling is fast growing in a young patient, he must be evaluated for Ewing's sarcoma as early diagnosis and treatment can lead to a better outcome and will prevent the metastasis.

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