Abstract: Rosai dorfman's disease is otherwise known as sinus histiocytosis with massive lymphadenopathy is a rare, idiopathic, benign disorder of reticuloendothelial system that causes painless cervical lymphadenopathy. The extranodal manifestation of the disease which is very rare. Here we report a similar case of 26 year old male with right infratemporal fossa mass extending to lateral orbit, which on radiological investigations came to differential diagnosis of Lymphoma, Granulomatous lesion, Schwannoma, Tuberculosis, Sarcoidosis. All speciality opinion were inconclusive and no improvement on medical management. Then we decided for the excision of the mass through an external approach. The post operative HPE report came as Rosai Dorfman Disease.

Key word: Rosai-Dorfman Disease, sinus histiocytosis, emperipolesis, extranodal, pterygopalatine fossa

INTRODUCTION

ROSAI DORFMAN’S DISEASE (SINUS HISTIOCYTOSIS WITH MASSIVE LYMPHADENOPATHY)

Sinus histiocytosis with massive lymphadenopathy (SHML) is a rare, idiopathic, benign disorder of reticuloendothelial system that causes painless cervical lymphadenopathy. It is classified as one of the histiocytoses, the proliferating cell is an antigen processing histiocyte. The clinicopathological entity was first described in 1960s by various investigators including Lemertin in 1964 and Azoury and Reed in 1966. Later, Rosai and Dorfman published additional cases and named the disorder that has become known as Rosai-dorfman’s disease or syndrome. It has also been called Destombes-Rosai –Dorfman’s syndrome. Accompanying the lymphadenopathy, which can be gigantic, with fever and certain laboratory abnormalities. The disease may affect extra nodal locations, and these tend to be at various head and neck sites. The disease is usually insidious, tends to be persistent or recurrent and has variable clinical course in each patient. It can cause significant morbidity and even death, usually a result of invasion of vital structures or from overwhelming infection secondary to immune dysfunction. Male : female ratio is 2:1. This disease is probably underdiagnosed, and because of paucity of cases there is typically a delay in diagnosis. The prognosis is usually excellent except in about 10% of patients. Those with extensive nodal disease, immunologic or hematologic abnormalities, involvement of unusual sites or multisystemic involvement have poor prognosis.

CASE REPORT

HISTORY AND EXAMINATION
26 year old male, came with complaints right frontal headache for 1 month Right eye defective vision-(near & distant vision) and double vision -1 month; Ear, Nose, Throat

On examination of Right eye the EOM was full. There was only perception of light with mild exophthalmos and temporal disc pallor. Relative Afferent Pupillary Defect (RAPD) present. Corneal reflex was intact. Left Eye examination was normal.

INVESTIGATIONS

Basic blood investigations, chest X ray, ECG were normal. CT-PNS : All sinuses were normal except for mild mucosal thickening.

CT Angiogram

Less vascular mass occupying pterygopalatine fossa, infratemporal fossa.
Radiologist gave differential diagnosis as Minor salivary gland tumor with perineural spread /Lymphoma/Granulomatous lesion.

Erosion of posterosuperior wall of right maxillary sinus, heterogeneously enhancing mass in the right pterygopalatine fossa extending through pterygomaxillary fissure to infratemporal fossa. Radiological differential diagnosis-Schwannoma/tuberculosis/sarcoidosis.

FURTHER MANAGEMENT

All specialty opinions were inconclusive about diagnosis. Biopsy attempted with DNE but no mass seen inside the maxillary sinus. Patient was given 2 weeks oral corticosteroid therapy but there was no improvement. Then we decided for excision of the mass by an external approach. Findings from CT & MRI shows the mass extension as follows. Infratemporal fossa—Pterygomaxillary fissure—Infraorbital foramen—Lateral orbit

SURGERY

Lateral Fisch Type D2 approach was done under General Anesthesia and tumor removed.
The symptoms and signs depend on the location and extent of disease. Nasal obstruction, epistaxis, saddle nose deformity and recurrent sinusitis can result from nasal and sinus involvement. Proptosis, ptosis, and decreased visual acuity can result form orbital, lacrimal gland, and base of skull disease respectively. Most symptoms are related to mass lesions in critical areas-for instance, ptosis, and facial pain in patients with lesions in pterygo palatine fossa. Between 30% and 40% of patients have extra nodal disease, commonly in the head and neck region. The nasal cavity, paranasal sinuses, orbit, salivary glands, gingival, buccal mucosa, facial bones, facial skin, trachea, larynx, thyroid, tongue, base of skull, temporal bone, nasopharynx, infra temporal fossa, and pterygo palatine fossa have all been reported to be sites of involvement.

Lesions in the subglottis are relatively common and can appear as polypoid, nodular, or exophytic masses. These cause symptoms of hoarseness cough and often stridor. The subglottic area and upper trachea. Once the airway has been secured, direct laryngoscopy and sharp excision or carbon dioxide laser ablation can be used. External beam radiation is not required for those with lesions affecting the larynx and involving upper aerodigestive tract. A tracheostomy is often required for those with lesions affecting the larynx and upper trachea. Once the airway has been secured, direct laryngoscopy and sharp excision or carbon dioxide laser ablation can be used. External beam radiation is not helpful; however, some chemotherapeutic agents have been used with some success including cyclophosphamide, chlorambucil, and etoposide. Recurrence often occurs, although eventually the disease regresses and becomes stable. Therefore, some practitioners recommend intervention solely for purpose of diagnosis or relief of symptoms. There are several treatment options, and therapy should be dictated by clinical situation. In many cases, observation is sufficient except for CD1a. Electron microtomic microscopy will show cytoplasm with lipid vacuoles and lysosomes and absent Birbeck granules.


HISTOPATHOLOGICAL EXAMINATION

A) Fibrotic capsule
b) Histocytes had vesicular nuclei, distinct nucleoli & abundant pale cytoplasm

c) Emperipolisis- Lympho&granulo phagocytosis HPE Report came as

ROS AI D ORFMAN DISEASE. POST OPERATIVE IMAGE

Minimal subconjunctival hemorrhage& restriction of mouth opening,no restriction of extracocular movements.

DISCUSSION

CLINICAL FEATURES:
The symptoms and signs depend on the location and extent of disease. Nasal obstruction, epistaxis, saddle nose deformity and recurrent sinusitis can result from nasal and sinus involvement. Proptosis, ptosis, and decreased visual acuity can result from orbital, lacrimal gland, and base of skull disease respectively. Most symptoms are related to mass lesions in critical areas-for instance, ptosis, and facial pain in patients with lesions in pterygo palatine fossa. Between 30% and 40% of patients have extra nodal disease, commonly in the head and neck region. The nasal cavity, paranasal sinuses, orbit, salivary glands, gingival, buccal mucosa, facial bones, facial skin, trachea, larynx, thyroid, tongue, base of skull, temporal bone, nasopharynx, infra temporal fossa, and pterygo palatine fossa have all been reported to be sites of involvement.

Lesions in the subglottis are relatively common and can appear as polypoid, nodular, or exophytic masses. These cause symptoms of hoarseness cough and often stridor. The subglottic area and upper trachea is also narrowed circumferentially. Because there may be midline osseous, cartilaginous, and soft tissue destruction and necrosis, SHML should be included in the differential diagnosis of midline destructive lesions. Other extra nodal sites include the external genitalia, the vertebrae, lungs, retro peritoneal structures (kidneys, pancreas, and so on) and the CNS. Most patients also have nonspecific symptoms, such as fever, weight loss, and malaise. Some patients have associated clinical syndromes, such as amyloidosis, wiskott-alrichs syndrome, and arthritis.

INVESTIGATIONS:
Laboratory studies often show anaemia, neutrophilic leucocytosis, an increased erythrocyte sedimentation rate, and polyclonal hypergammaglobulinemia. Radiographic studies are often performed but are usually only helpful if extranodal disease is present. The differential diagnosis depends on the location of the lesion and the list can be long. For instance Langerhans’ cell histiocytes, inflammatory pseudotumour, plasma cell granuloma, rhinoscleroma, Gaucher’s disease, extra medullary plasmacytoma, neurofibromatosis, lipid storage diseases, metastatic malignancy, and many others may have to be considered depending on clinical history. HISTOPATHOLOGICAL EXAMINATION Histopathological specimens characteristically show dilated lymph node sinuses filled with large histiocytes that contain intracytoplasmic lymphocytes, plasma cells, and neutrophils. The lymphocytes are B cells, T cells, and natural killer cells. This process of cytophagocytosis is called emperipolisis, which is very characteristic of Rosai-Dorfman’s disease. The medullary cords may have numerous plasma cells. The lymph nodes increase in size and have a thick fibrous capsule. There are hyperplastic germinal centres but not an increased number. Large histiocytes are present with round nuclei, prominent nucleoli, and abundant pale cytoplasm. The presence of lymphocytes, neutrophils, and plasma cells is also noteworthy. The histopathology of extranodal disease is interesting. Dark staining areas alternate with pale foci. The dark areas are aggregates of lymphocytes, and the pale areas are foci of histiocytes with some lymphocytes and plasma cells. Typically, fibrotic bands are present, similar to fibrotic nature of lymph node capsule, and there tends to be less emperipolisis. Special stains & Immunohistochemistry - S – 100 protein, PAS, CD 68, CD 14, HAM 56 NEGATIVE for CD1a. Electron microtomic microscopy will show cytoplasm with lipid vacuoles and lysosomes and absent Birbeck granules.

The characteristic pathologic findings, particularly emperipolisis, eliminate all other diagnostic possibilities except LCH (Langerhan Cell Histiocytosis). LCH can be ruled out by electron microscopy. TREATMENT There are several treatment options, and therapy should be dictated by clinical situation. In many cases, observation is sufficient as long as there is no systemic involvement. A bone marrow biopsy, thorough physical examination and routine laboratory studies are required. Surgical intervention may be mandated in certain situations. Endoscopic sinus procedures can be performed to prevent intracranial extension and to treat recurrent or chronic sinusitis. Airway management problems occur in patients with disease involving upper aerodigestive tract. A tracheostomy is often required for those with lesions affecting the larynx and upper trachea. Once the airway has been secured, direct laryngoscopy and sharp excision or carbon dioxide laser ablation can be used. External beam radiation is not helpful; however, some chemotherapeutic agents have been used with some success including cyclophosphamide, chlorambucil, and etoposide. Recurrence often occurs, although eventually the disease regresses and becomes stable. Therefore, some practitioners recommend intervention solely for purpose of diagnosis or relief of symptoms. There are several treatment approaches to infratemporal fossa (ITF) viz, preauricular (subtemporal), postauricular (transtemporal), Lateral Fisch Approach, Anteriortransfacial Approach(Facial Translocation), Transorbital Approach. Lateral Fisch Approach has 4 types Type B & C- more anterior pathology involving petrous apex & dural Type C is for lesions of anterior ITF, sella, nasopharynx. Hallmark of these approaches include facial nerve rerouting, subtemporal/dural exposure Type D - The Fisch type D exposure is a preauricular modification of the Fisch infratemporal approach. Type D1 addresses tumors of the anterior infratemporal...
fossa, whereas type D2 is designed for lateral orbital wall lesions and high pterygopalatine fossa tumors. The distinguishing feature of the type D approach from type B and C approaches is that the middle ear and eustachian tube area is not obliterated, and conductive hearing is not sacrificed. In addition, the infratemporal facial nerve is not rerouted, and the petrous internal carotid artery is not fully exposed. Although these preauricular approaches do not include a temporal craniotomy, the floor of the skull base can be drilled away to allow full access of infratemporal fossa. In our case we did right orbitozygomatic and removal of tumor without entering middle cranial fossa Fisch type D2. Some cases resolve spontaneously. It has 2% chances of recurrence. Rosai Dorfman disease is a very rare tumor difficult to diagnose preoperatively & can occur anywhere in the body and operated if symptomatic. Medical Treatment was not satisfactory in our case.

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