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RARE PRESENTATION OF SINONASAL MASS: EXTRA-NODAL ROSAI DORFMAN DISEASE: A CASE REPORT

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ABSTRACT

Introduction: Rosai -Dorfman disease (RDD) is a rare, usually self-limiting, benign condition which is characterised by over production and accumulation of a histiocytes in the lymph nodes resulting in painless cervical lymphadenopathy. Extra -nodal RDD is uncommon and have been reported in approximately 43 % of cases and can affects skin, respiratory tract, a solitary bone, nasal cavity, or the central nervous system. Histopathological examination plays an important role in diagnosis which shows emperipolesis. Definitive treatment yet to established due to its rarity and propensity for spontaneous remission. **Case report:** We reported a case of extra-nodal RDD involving the sinonasal area with intracranial extension without lymphadenopathy in a 55 years old male who presented with complaint of nasal obstruction with difficulty nasal breathing and later was diagnosed of Rosai Dorfman disease on histopathological examination. Patient was managed by surgical excision and adjuvant treatment with oral steroid and methotrexate. **Conclusion:** Due to lack of pathognomonic clinical and radiological diagnostic features it is difficult to diagnose RDD involving sinonasal area hence it is important to keep RDD as one of differential diagnosis in benign sinonasal mass. Histopathology examination plays a very important role in definitive diagnosis of RDD. Surgical intervention is only recommended when there is cosmetic deformities and obstructive symptoms. Long-term follow up is necessary to recognise and prevent the recurrences of disease.

KEYWORDS

Rosai Dorfman disease (RDD), histocytes, emperipolesis.

INTRODUCTION

In 1969, pathologists, Jaun Rosai and Ronald Dorfman first described the Rosai Dorfman disease (RDD) as sinus histiocytosis with massive lymphadenopathy and characterized by marked, painless lymph node enlargement [1,2]. It is a benign, self-limiting disease of unknown aetiology with spontaneous remission course.[1]However, some studies suggest that it is due to aberrant response to an unspecified antigen, possibly an infectious organism[9]. According to the extent of involvement, RDD can be classified as lymph node, extra-nodal, or mixed (i.e., simultaneous involvement of lymph nodes and extra-nodal organs). Although peripheral lymphadenopathy is the most common mode of presentation, numerous studies and individual case reports have established that 43% of affected individuals have extra-nodal manifestations[6].Among the extra nodal sites, the head and neck region is most commonly affected[8]. Most of the extra nodal cases of RDD in the head and neck region affect the nasal cavity, pharynx and paranasal sinuses. Other extra-nodal sites being skin, respiratory tract, a solitary bone, genitourinary system or the central nervous system. [2,3,7] The disease is diagnosed by biopsy of affected tissues and histopathological hallmark shows emperipolesis[5].Treatment options include corticosteroids, surgery, radiation therapy and chemotherapy[4].

Surgical excision is rarely advised until the disease has grown to an unmanageable size and is causing respiratory impairment or a cosmetic deformity.[10]

Case Report

A 52 years old male presented in our outpatient department with complaints of bilateral nasal obstruction for 5 years which is insidious in onset, slowly progressive, initially on left, progressed to right side which does not get relived with medications.

It was associated with anosmia, left hemi-cranial headache, mouth breathing and nasal twang. Patient also complain of bilateral reduced hearing. Patient denied any history of local pain, fever, weight loss or night sweats. Patient was of average built. There was no history of diabetes, tuberculosis or hypertension. History of previous nasal biopsy suggestive of only inflammation. No evidence of malignancy.

On examination there was a lobular mass occupying the whole left nasal cavity on anterior rhinoscopy. On probing there was no bleeding from the mass. Air-blast was absent on left side on cold spatula test. On examination of oral cavity and oropharynx there was bulge seen on the soft palate. Posterior rhinoscopy finding was normal. On otoscopic examination, bilateral tympanic membrane was normal. On tuning forks test, Air conduction was better than Bone conduction in all the three frequencies.

Visual acuity and extra-ocular movements were normal. There was no evidence of cervical lymphadenopathy on neck examination.

A CT scan of nose, paranasal sinuses and oropharynx was done & it was suggestive of 9x5.4x3.5 cm lobulated enhancing soft tissue lesion in left maxillary sinus, left choana and left nasopharynx with significant obliteration of left nasal cavity. Lesion extending into left pterygopalatine fossa with infiltration into left infratemporal fossa and masticator space. Infiltration of left orbital apex and fissure seen. Intra-cranial extension seen in middle cranial fossa, left cavernous sinus and temporal region.

Opacification of bilateral sphenoid and ethmoid sinuses. Right maxillary opacification seen. The nasal septum was deviated towards left. (Figure 1a, b, c, d)

MRI Brain with paranasal sinuses was done for better soft tissue delineation. (Figure 2 a, 2b)

Laboratory investigations revealed low haemoglobin (Hb=9.5g%) however, erythrocyte sedimentation rate (ESR), leucocyte count, neutrophil count, platelet count and eosinophil count were all in normal range. Serology tests (HIV, hepatitis -B and C) were non-reactive. X-ray chest was within normal limit.

Pure tone audiometry suggestive of right ear mild sensorineural hearing loss (32 DB) and left moderately severe sensorineural hearing loss (66 Db).

Patient was posted for nasal mass biopsy under general anaesthesia for confirmation of diagnosis. Maximum debulking of mass from left nasal cavity, sinuses, choana and nasopharynx was done and tissue was sent for histopathological examination.

Post-operative period went uneventful. Patient was transfused with two pints of whole blood to build up haemoglobin.

Histopathological examination revealed diffuse sheets of histiocytes

focally exhibiting emperipolesis and interspersed with plasma cells rich infiltrate. Patchy sclerosing fibrosis seen. A focal increase in IgG4 positive plasma cells was noted. However, IgG4/IgG ration is less than 40%. Histologic features specific for IgG4 related disease not seen. No granulomas seen. No malignant cells were seen. Overall features were consistent with Rosai-Dorfman disease. (Figure 3)

The patient was given chemotherapy(methotrexate), systemic oral corticosteroid(prednisolone), concurrent with iron and folic supplements. Patient was following up after 6 months with repeat MRI Brain and Para nasal sinuses which show near complete disease free from nasal and nasopharyngeal areas. However, intra-cranial part showed significantly increase in size. Patient was then referred to Neurosurgery and Haematology doctor for further treatment.

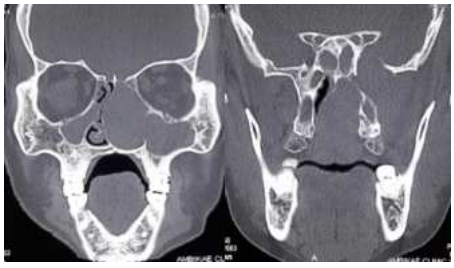

Figure 1a. CT PNS with brain in coronal view

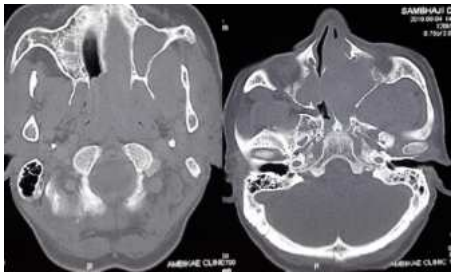

Figure 1b. CT PNS with Brain in axial view

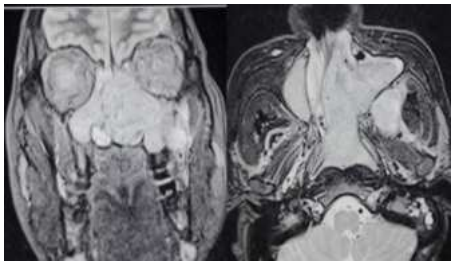

Figure 2a: MRI PNS with Brain in coronal and axial views

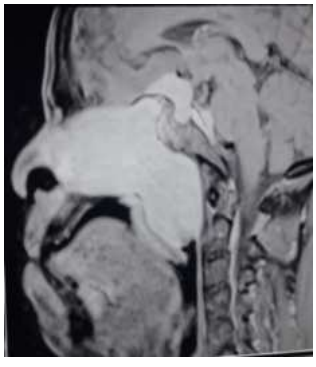

Figure 2b.MRI Brain with PNS in sagittal view

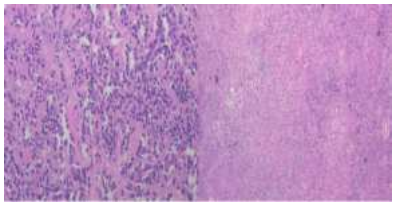

Figure 3. Histopathological examination showing prominent histiocytic infiltration in the background of inflammatory cells (plasma cells and lymphocytes) exhibiting Emperipolesis.

DISCUSSION

Rosai -Dorfman disease (RDD) is a rare, usually self-limiting, benign condition characterised by over proliferation of histiocytes[1,2]. It mainly affects the lymphatic tissues[7] However, the disease may manifest in extra-nodal sites as well.

It was first described by Jaun Rosai and Ronald Dorfman in 1969[1].The classical presentation of RDD is bilateral painless massive cervical lymphadenopathy with or without fever, night sweats and weight loss[1,2].

The extra-nodal RDD involvement is uncommon and have been reported in 43% of cases and most commonly affects skin, nasal cavity, respiratory tract, a solitary bone, or the central nervous system. [3,6,7,8] The aetiology remains unknown. However, some studies suggest that it is due to aberrant response to an unspecified antigen, possibly an infectious organism by Ebstien Barr virus or klebsiella.[9] Histopathological examination along with immunohistochemistry plays an important role in definitive diagnosis of RDD. The pathognomonic histopathological feature is emperipolesis Which is a prominent histiocytic infiltrate in a background of inflammatory cells, predominantly comprised of lymphocytes and plasma cells [5]. The histopathologic features of RDD in extra-nodal sites were found to be similar to nodal disease except fibrosis tends to be more pronounced and emperipolesis less evident. Immunohistochemical features usually shows CD 68 and CD 1a positivity, positive staining for S-100 protein.[12,13]

The haematological picture usually demonstrates anaemia, leucocytosis, neutrophilia and increased ESR.[2]

Radiological imaging is necessary for better understanding of disease extension and any intracranial extension especially in an extensive disease.

In our case, immunohistochemistry study was not done and laboratory examination revealed anaemia.

The 70%–80% of patients recovered from symptoms without treatment. The standard treatment protocol for RDD is yet to be determined due to rarity of the disease and its propensity for spontaneous remission.[11]

Excision, radiation therapy, chemotherapy and systemic steroids have all been tried by different authors.[4] However, their efficacy has not been established.

In our case, patient was managed by endoscopic excision and adjuvant therapy like methotrexate and systemic corticosteroids. Follow up after 6 months with repeat MRI of brain and PNS shows near complete free of disease from nasal and naso-pharyngeal areas however intracranial component shows significantly increase in size. Patient was then referred to Haematology and Neurosurgery doctor for further management.

CONCLUSION

- Due to lack of pathognomonic clinical and radiological diagnostic features it is difficult to diagnose RDD involving sinonasal area hence it is important to keep RDD as one of differential diagnosis in benign sinonasal mass.
- Histopathological examination plays a very important role in definitive diagnosis of RDD.
- Surgical intervention is only recommended when there is cosmetic deformities and obstructive symptoms.
- Long-term follow up is necessary to recognise and prevent the recurrences of disease.
- More study needed to form a definitive management protocol.

Conflict of Interest: - There is no conflict of interest

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