SOLITARY PAROTID MASS PRESENTING AS SARCOIDOSIS: UNUSUAL PRESENTATION

*V. V. Rokade, **N. A. Pathak, ***S. V. Nemade

ABSTRACT:

Objective: To demonstrate an unusual case of sarcoidosis in which the patient presented with solitary parotid mass and no other manifestations of the systemic disease.

Case report: A 12 years old girl child presented with history of swelling in right parotid region since three years. On local examination there was firm smooth non tender swelling palpable in right parotid region. FNAC was suggestive of chronic granulomatous disease. Sr Calcium, ACE enzymes, Serology, ANCA was normal. Monteux test was negative. ECG was normal. Superficial parotidectomy was done. Histopathology features were suggestive of sarcoidosis.

Conclusion: We believe that this case is of interest as descriptions of such presentations of sarcoidosis as a solitary discrete mass in parotid gland and absence of systemic involvement are not prevalent in the literature. As all the investigations were normal, only histopathology report after superficial parotidectomy guided us to reach up to final diagnosis of sarcoidosis.

Keywords: Parotid Sarcoidosis, Non caseating granulomatous disease.

INTRODUCTION:

Jonathen Hutchinson first reported sarcoidosis in 1869. Several causative agents have been implicated in its aetiology which includes infective agents like mycobacterium and no infective agents like exposure to beryllium dust. Detection of mass of parotid gland is generally considered an indication for superficial parotidectomy. In most cases pathology will identify a euplastic process. We describe a case in which patient presented with discrete solitary parotid mass that was turned out to be sarcoidosis on histopathology.

CASE REPORT:

A 12 years old girl child presented in ENT OPD with history of swelling in right parotid region since three years with no history of fever or fluctuations in size with or without eating. General practitioner treated her with antibiotics with no improvement.

On local examination there was firm smooth non tender swelling palpable in right parotid region. Duct opening was normal. There were no signs of facial paralysis. General examination revealed no evidence of peripheral lymphadenopathy & no hepatosplenomegaly. CVS, RS and CNS examination were within normal limits.

On diagnostic work up her haemogram and ESR was normal. FNAC showed plenty of epithiloid cell granulomas with multinucleated giant cells; features suggestive of chronic granulomatous disease.

Corresponding Address:
Dr. Vidya V. Rokade,
A-2, 303 Sun Empire, Sun City Road
Vadgaon Bk., Pune-51
Ph.: 9922160881
E-mail: vidyarokade@hotmail.com
To rule out various granulomatous diseases various investigations carried out. Biochemical investigations: Blood Sugar, KFT, LFT, Sr Calcium, ACE enzymes, Serology, ANCA were normal. Montex test was negative. ECG was normal. Radiological investigations: Xray chest, USG abdomen revealed no abnormality. USG parotid with colour Doppler showed enlarged right parotid gland with multiple lymph nodes in it.

CT& MRI (plain & contrast) demonstrated enlargement of right parotid gland with homogenous enhancement with involvement of superficial lobe of gland with multiple lymph nodes in it (FIG I).

Inspite of long list of investigations we could not arrive at the final diagnosis. Hence, to obtain tissue diagnosis superficial parotidectomy was done. Intraoperatively we found firm mass in superficial lobe of parotid gland with two to three enlarged lymph nodes. Deep lobe was normal on palpation. Histopathogy showed numerous non caseating epitheloid cell granulomas surrounded by lymphocytes & Langerhan's type of giant cells. Also seen were Schwann's bodies in giant cells & asteroid bodies as well as Hamazaki Wegenbreng inclusions with no evidence of malignancy. Z N staining was negative for AFB. All the features were suggestive of sarcoidosis (Fig II).

Postoperative period was uneventful. Patient is under close follow up for last one year without any medical treatment. Her x ray chest remained normal and had not manifested any symptoms of systemic sarcoidosis.

**DISSCUSSION:**

Despite extensive studies, no agent has been identified as the cause of sarcoidosis. Sarcoidosis is currently considered as a chronic inflammatory disease, distinguished by hyperimmune reactions to an unspecified agent at the lesion sites.¹

Sarcoidosis affects lungs in 90% of cases followed by lymph nodes & spleen. Salivary glands are rarely involved.

Parotid gland involvement occurs in 0.5 to 15% patients of systemic sarcoidosis.² However, in 6% of patients it is confined to parotid gland only & may

---

¹ For a detailed explanation of sarcoidosis, refer to the relevant section in the source material.

² For further details on parotid gland involvement, consult the additional sections in the source.
manifest as uni or bilateral painless swelling. Parotid enlargement is a more frequent finding in children diagnosed with early onset sarcoidosis.

Parotid gland involvement in sarcoidosis may manifest itself in a variety of clinical patterns. The most common pattern is represented by bilateral diffuse enlarged parotid gland. A second pattern is characterized by asymptomatic parotid lesions. Third pattern is in the form of Hereford’s syndrome. A Clinical pattern of sarcoidosis characterized by isolated parotid enlargement with absence of general symptoms of sarcoidosis would be extremely uncommon.

In cases of solitary parotid sarcoidosis preoperative clinical diagnosis is difficult as there is no specific lab test as such. However FNAC can be helpful. Depending upon the conclusion of FNAC we have to rule out various infective and granulomatous conditions. Markers of activity for sarcoidosis include raised serum ACE levels, abnormal calcium metabolism, positive Kveim Slitizbach skin test & radioactive gallium scanning (Gallium-67 Citrate). However, ACE levels are typically elevated in 80% of children and 60% of adults with late onset sarcoidosis. The ACE test has proven to be less sensitive in patients diagnosed with early onset sarcoidosis. Many other conditions are associated with elevated ACE levels like milliary TB, Leprosy and Gaucher’s disease. It is important to rule out various infectious and granulomatous diseases on the basis of history and physical examination, serologic testing and special tissue staining techniques. The combination of an elevated ACE levels and positive gallium-67 scan is 85% specific for sarcoidosis.

A definitive diagnosis of sarcoidosis is best achieved by integrating clinical data with presence of noncaseating granulomas. The main goal of the treatment is to minimize or prevent inflammation & granuloma formation that ultimately causes end stage organ destruction by hyaline fibrosis. Steroids remains the cornerstone of treatment for symptomatic patients and those with the progressive disease, although the dosage and the duration of the treatment are not well defined and no consensus exists as to whether treatment should be given to asymptomatic patients and patients with only mild disease. Patients are usually treated for at least six months initially.

Methotrexate. Azathioprine, Cyclophosphamide & recently Infliximab has been tried with varying success rate for the patients refractory to steroids.

However, spontaneous regression of this disease has been reported. Early diagnosis and treatment can slow up or stop the progression to organ destruction. Research is still on going regarding better diagnostics & treatment modalities.

SUMMARY:

- We believe that this case is of interest as descriptions of such presentations of sarcoidosis as a solitary discrete mass in parotid gland and absence of systemic involvement are not prevalent in the literature.
- As all the investigations were normal, only histopathology report after superficial parotidectomy guided us to reach up to final diagnosis of sarcoidosis
- So we believe this makes the case an unusual presentation.

Acknowledgement: We would like to express our appreciation to Dr S.D.Deshmukh Professor, Department of Pathology SKNMC, Pune for prompt and accurate diagnosis.

DISCLOSURES

(a) Competing interests/Interests of Conflict - None
(b) Sponsorships - None
(c) Funding - None
REFERENCES:


