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RESEARCH ARTICLE

A RARE CASE OF CHRONIC APPENDICITIS WITH ROSAI DORFMANS DISEASE OF MESENTERY.

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Abstract

Appendicitis is a global epidemic with 72,000 deaths globally in 2013 less in number than from 88,000 in 1990.[1]Rosai Dorfman disease i.e. sinus histiocytosis with massive lymphadenopathy, is a rare disorder of unknown etiology that is characterized by abundant histiocytes in the lymph nodes throughout the body[2,3].Lymphadenopathy in the neck is commonest site for occurrence however it is rarely associated with mesentry.[4] When Appendicitis secondary to Rosai Dorfamns disease is there it turns out to be the rarest one. Here reporting a same case Appendicitis with Rosai Dorfaman Disease In a young Female patient.

Introduction:-

In 1969, two pathologists, Juan Rosai and Ronald Dorfman, reported a distinct histiocytic disorder in several patients with massive enlargement of the lymph nodes, as well as other symptoms. They named this condition sinus histiocytosis with massive lymphadenopathy, and the name has since come to be known as Rosai-Dorfman disease.Rosai Dorfaman disease is the rarest histiocytic proliferative disorder of unknown aetiology, also known as Sinus Histiocytosis with massive lymphadenopathy.[5,6] Classically RDD usually presents as massive, painless, enlarged bilateral neck lymph node, approximate 40% of RDD cases documented to date present in extra nodal organs or tissues, in some without associated lymphadenopathy which may or may not develop later in the disease course.[7,8] Gastrointestinal tract involvement of Rosai Dorfman disease is very rare with only 1% of extra nodal manifestation of the same recorded until today.[9,10] Only few reported cases are there in literature having Appendicitis with Rosai Dorfmans disease.[11]

Case report:-

A female patient of age 25 years residing in Pune presented with the complaints of severe pain at right iliac fossa, fever on and off, cold and coryza. Patient has taken treatment in other hospital and visited to current hospital with USG report of her and was previously diagnosed as Acute Appendicitis.

Patient has no history of vomiting, nausea, swelling in any part of body, weight loss and general systemic examinations were normal. Local examinations includes per abdomen palpation having tenderness in right iliac fossa, cough reflex is positive, rebound tenderness positive, heamogram shows leucocytosis with Alvarado scale[12]8. Last menstrual period was ten days back of IPD admission date.

Laboratory investigations suggest leucocytosis(11,000) with 1-2 epithelial cells in urine routine examination and negative serological report. Ultra Sonography (A/P) shows significant probe tenderness noted in RIF, underlying lesion shows small hypo echoic lesion measuring 16x16mm.

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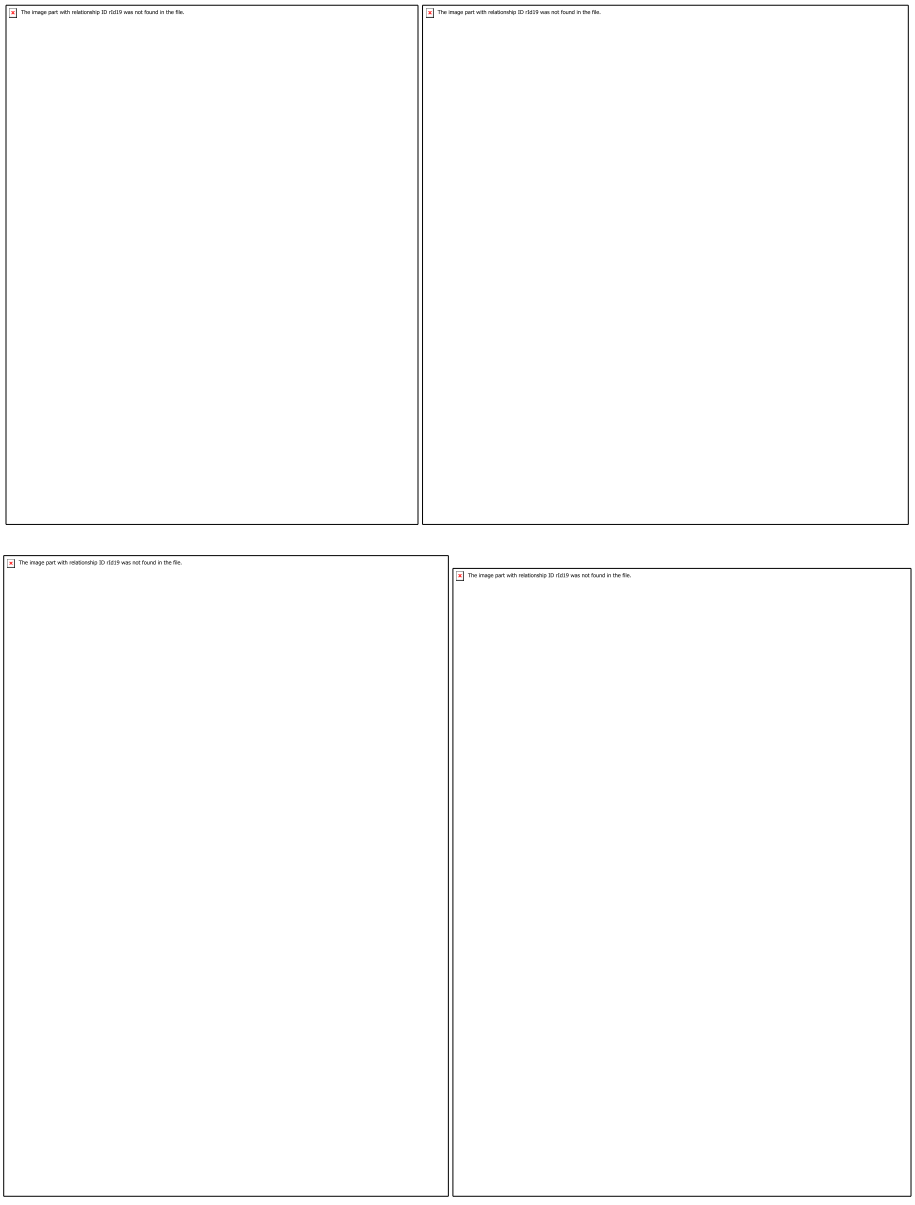
After thorough clinical examinations decision was taken to treat appendicitis. Hence appendectomy was decided. Prior to surgery written consent and physicians fitness was taken. Two diagnoses were kept in mind, first was acute appendicitis as Alvarado scale was high, secondary any neoplastic lesion for which biopsy was needed.

For both decision, laparotomy was needed and patient was operated for appendectomy. Patient was operated for appendectomy through Grid Iron incision. Inflamed appendix was removed and a lesion of 2x2x2cm at anterior, medial to tinea coli near to ileo-cecal joint was firm but not hard, full thickness biopsy was taken and simple tube drain was kept in right Para-colic gutter. Specimen of lesion, appendix and two mesenteric lymph node was sent for histopathology. Diagnosis was perforated ileocaecal junction with peritonitis, fat necrosis, and peri ceacal abscess with chronic appendicitis and Sinus histiocytosis. Treatment include antibiotics from cephalosporin group, analgesic, antacid for seven days by intra venous route and further by oral route.

Patient was discharged on seventh day with normal gastro-intestinal function. On fifteenth day stiches were removed and patient was reviewed for ultra sonography after two months which shows normal reports.



Disscussion:-

Rare histiocytic disorders which involve over production of a type of white blood cell called non Langerhans's sinus histiocyte. These cells then accumulate a most often a lymph nodes but may occur in other area of body and can lead to organ damage. The reason of over production is not known, although many possibilities have been considered.

In approximately 43% of cases in other areas of body besides lymph nodes may also be affected. Some individuals may have extra nodal disease without presence of lymphadenopathy. The symptoms and physical finding associated with Rosai Dorfman Disease vary greatly from one person to another.

Treatment in many cases, the symptoms of Rosai Dorfman disease may disappear without treatment (spontaneous remission) within in months or a few years. Clinical observation without treatment is preferred for individuals with Rosai-Dorfman disease whenever possible. In many cases, no therapy will be necessary.

In some cases, various treatment options may become necessary. In these cases, the treatment of Rosai-Dorfman disease is directed toward the specific symptoms that are apparent in each individual. Several different treatment options have been used to treat individuals with Rosai-Dorfman disease including surgical removal of histiocytic lesions. In more serious cases, additional treatment options have included therapy with certain drugs including steroids (e.g., prednisone), alfa-interferon, and a regimen of certain anticancer drugs (chemotherapy). In some cases, affected individuals have shown improvement of symptoms with these treatments. In other cases, drug therapies have been ineffective. Other treatment is symptomatic and supportive.

Conclusion:-

In above case surgical removal of lesion was carried and follows up was taken upto six month. Abdomen sonography does not show any evidence of reoccurrence.

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