Xanthogranuloma of Abdominal Wall - A Rare Case Report

Ruchi Khadayate¹, Reeta Dhar², Siddhi Karale¹

¹Second year Post- Graduate student, ² Professor, Department of Pathology, MGM Medical College & Hospital, Kamothe, Navi Mumbai
Corresponding Author: Reeta Dhar

ABSTRACT

Xanthogranulomatous inflammation is a rare inflammatory process characterized by proliferation of histiocytes and Touton giant cells. It can simulate a neoplastic process. The causative agent can be any infectious process or any causative organism, inflammatory conditions or lysosomal storage disorders like Niemann-Pick, Gaucher’s disease etc. Xanthogranulomatous inflammation comes in the category of Non-Langerhans cell histiocytosis. Pathologists should know the histologic features of this histiocytic disorder to categorize into and also help in patient management.

Keywords: Xanthogranuloma, Abdominal wall, Non-Langerhans cell histiocytosis

INTRODUCTION

Xanthogranuloma is the most common type of Non-Langerhans cell histiocytosis. It is caused by proliferation of histiocytes (macrophages). Two of the most important Histiocytic disorders are Juvenile Xanthogranuloma and Langerhans cell histiocytosis. Juvenile Xanthogranuloma is a disease of infancy or childhood and most commonly presents as cutaneous nodules over head and neck region. Xanthogranulomas occur in adult life also with involvement of deep soft tissue, testis, pericardium, lung, kidney, retroperitoneum and spleen. (¹)

CASE REPORT

A 71 yrs old male patient came to casualty with abdominal pain, distension and difficulty in passing urine and stools since 4 days. Patient is chronic alcoholic and smoker since past 15 years. Patient was admitted under surgery and full workup of the patient was done. The complete blood count revealed Hb- 9.8gm/dl, TLC- 35000cells/cumm, Plt-1.90 lakhs/cumm. On USG abdomen, free fluid in perihepatic, perisplenic and interbowel pelvis with multiple septation was noted suggestive of Chronic Ascites. In view of radiological findings, patient was posted for Exploratory Laparotomy Surgery in view of Pyoperitoneum and Appendicitis. During Exploratory Laparotomy, Appendix with part of peritoneum and tissue connecting the anterior abdominal wall (umbilicus ) to the bladder(muscle tract)- was resected and sent to Histopathology section of the Central Laboratory of MGM Medical College, Kamothe, Navi Mumbai.

Gross Finding-
Received three specimens as follows-
1. Labelled as Appendix- Single, tubular, grey white to grey brown, soft to firm tissue piece measuring 6cm in length. External surface- congested.
   On cutting the tissue- Lumen identified.
2. Labelled as muscle tract- Multiple, grey brown, soft to firm tissue pieces largest measuring 3.5x2.5x1cm and smallest measuring 1cm in length.
3. Labelled as Omentum- Single, grey-yellow, soft to firm tissue piece measuring 3.5x1.0x0.5cm.
Microscopic Finding-

Appendix and Omentum revealed features of Acute Appendicitis (Transmural acute inflammatory infiltrate). No multinucleate giant cells or granulomas noted in the sections studied.

Histological features from the muscle tract revealed ill circumscribed, non-capsulated lesion comprising of predominantly fibrocollagenous tissue infiltrated by mixed inflammatory infiltrate composed of lymphocytes, plasma cells, histiocytes and numerous multinucleate Touton type of giant cells along with bundles of skeletal muscles and myofibroblasts. Few focal collections of Xanthoma cells and histiocytes with foamy to slightly eosinophilic cytoplasm are also noted. The infiltrate is seen extending to surrounding muscle and fat tissue. Focal collections of neutrophilic infiltrate with eosinophils and lymphocytes with congested blood vessels and hemorrhage are seen. No well formed epithelioid granuloma or caseous necrosis were noted. No lining epithelium or cystic structure identified in the sections studied.

Special stains with Ziehl-Neelsen, Fite-Faraco, Periodic Acid–Schiff, and Grocott Methenamine Silver Nitrate stains were negative hence no evidence of any Koch’s parasite or fungal infections were demonstrated. Diagnosis of Inflammatory lesion suggestive of Xanthogranuloma was given.

Figure 1- Micrographs show numerous multinucleate giant cells (Red arrows) along with collagenous tissue, skeletal muscle bundles (Green arrow) and inflammatory infiltrate. (H & E stain 4X)

Figure 2- Micrographs show numerous Touton type of multinucleate giant cells (Arrow head) along with numerous xanthoma cells (foamy macrophages (Arrows) and inflammatory infiltrate comprising of lymphocytes, plasma cells, eosinophils and neutrophils. (H&E stain 10X)
Figure 5 – Micrographs shows eosinophils (arrow) in the inflammatory infiltrate with fibroblastic proliferation. (H&E stain 10X)

Figure 6 - Micrographs shows areas of haemorrhages (arrow head) along with dilated congested blood vessel and foamy macrophages . (H&E stain 10X)

Figure 7- Micrographs shows Touton giant cell showing multiple nuclei forming a ring surrounded by foamy cytoplasm making the cytoplasm visible around the nuclei. And also seen are foamy macrophages and lymphocytes. (H&E stain 40X)

DISCUSSION

Histiocytes are considered to arise from stem cell precursors in the bone marrow that undergo differentiation toward the antigen-presenting cell (class I) or monocyte-macrophage (class II) lineage. (2) Xanthogranulomatous inflammation is a rare aggressive inflammatory conditions that can be caused by infection, inflammation, histiocytic processes, or inherited disorders [3,4] and are not pathognomonically specific but are characterized by admixture of granular, eosinophilic, periodic acid-Schiff–positive histiocytes, foamy macrophages, and plasma cells accompanied by a varying degree of supplicative inflammation, abscess formation, or hemorrhage. (4) There can be capillary proliferation, collagen formation, and hemorrhages. The histiocytes in close proximity to the hemorrhage engulf the erythrocytes (erythrophagocytosis), forming lipid-laden histiocytes (xanthoma cell). This inflammatory process results in granulation and necrotic debris. (5) This process eventually leads to replacement of the parenchymatous cells with abundant xanthoma cells, fibrin, collagen deposition and tissue destruction.

Xanthogranulomatous pyelonephritis, cholecystitis and of visceral organ and genitourinary organs are a very known entities. The causative mechanism is the extensive inflammatory response against the various mechanical and microbiological insults to the particular organ. They are known to involve the surrounding structures like the mesenteries simulating aggressive process leading to abscess formation mimicking a tumour mass radiologically. (6)

Various organs of the body are known to be involved with kidney and the gall bladder being the commonest ones. (7) Other predominantly involved sites, which have been reported are endometrium, epididymis, fallopian tubes, bone, skin, appendix, urinary bladder, thyroid and adrenal glands. (8,9) Spillage of gall stone or perforation of appendix or involvement of other visceral organs causes spread to the surrounding structures is a known cause of xanthogranulomatous inflammation of the abdominal cavities. Exact pathogenesis of
xanthogranulomatous inflammation is not yet known but different hypothesis are given like defective lipid transport, disorders of neutrophilic chemotaxis, lymphatic obstruction and specific immune response towards Proteus and E. coli. (10)

Systemic Xanthogranulomatous process is a rare disorder of non Langerhans cell histiocytosis. They are comprised of Erdheim-Chester disease (ECD), Rosai-Dorfman disease (RDD), hemophagocytic lymphohistiocytosis (HLH), and juvenile xanthogranuloma (JXG). These disease show multisystem involvement with diagnosis is made on radiological, histopathological and immunohistochemical findings. (11)

In our case, peritonitis was present but the appendix, gall bladder and omentum did not show xanthogranulomatous inflammation and also there was no evidence of perforation of the abdominal viscerae. Serous and pus collection was noted over the bladder with a tract like tissue from the bladder to anterior abdominal wall. No signs of obstruction were noted in bowel. All though Patient gave history of gastrointestinal ulcers for which he was operated 40 yrs back. But no current symptoms or relevant history was present.

CONCLUSION

We would like to conclude by saying that our case presented with intramuscular xanthogranulomatous inflammation which was mostly an acquired disorder with no evidence of xanthogranulomatous inflammation in the gall bladder, appendix or any other resected tissue. Intraoperative finding of the muscle tract connecting the anterior abdominal wall to bladder suspecting it to be urachus but no lining epithelium was seen in the multiple sections from the resected tissue.

REFERENCES


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