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Case Report

Extranodal (cutaneous) Rosai Dorfman: a rare presentation

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ABSTRACT

Rosai Dorfman disease is a rare histiocytic disorder of unknown etiology commonly involving the cervical lymph nodes first described in 1969, Extranodal involvement is very rare. Clinical course is variable.

Keywords: Extranodal, Rosai Dorfman

INTRODUCTION

Rosai Dorfman disease is a rare histiocytic disorder of unknown etiology commonly involving the cervical lymph nodes first described in 1969, since then more than 400 cases have been reported in the Rosai Dorfman disease registry. Extranodal involvement have been reported in about 20-40 % of cases which includes skin, soft tissue, upper respiratory tract, bones, eye, urogenital tract, breast, gastrointestinal tract and lung.[1,2] We report a case of extranodal involvement of the skin in a young male.

CASE REPORT

A 34-year-old male presented to the dermatology department with chest lesion approximately measuring 5x5 Cm size. It was treated outside as a keloid and was given intralesional injections few months back. On examination lesion was ulcerated and bleeded on touch. According to the patient the lesion was peanut size 8 months back and by now it has increased to this size. Laboratory investigations were done, counts were in normal limit except ESR was slightly raised. Abdominal USG was done and showed no evidence of any hepatomegaly or spleenomegaly. No axillary or neck nodes were noted. Surgery was planned, and lesion was removed after consent, patient was sent home and undergoing followup with no Chemotherapy. On histopathological examination microscopy of the specimen revealed stratified keratinized epithelial layer below dermis showed plasma cells, neutrophils, histiocytes and large cells with vesicular nuclei and abundant cytoplasm exhibiting emperipolesis consistent with diagnosis of Rosai Dorfman Disease. IHC markers were done, S100 positive and CD1A negative.

DISCUSSION

This condition was first described by Robb-Smith in 1947 in children and was termed as giant cell sinus reticulosis.[3] Sinus histiocytosis with massive lymphadenopathy (SHML) hasbeen recognized as a distinct clinicopathological entity, though first given this name by Rosai and Dorfman in 1969, 1972. The predominant clinical manifestation of the disease is tumoral cervical adenopathy (87.3% of cases) that, in most cases, is painless and bilateral, affecting one or all cervical chains. The neck lymph nodes are the most frequently involved, followed by inguinal, axillary and mediastinal lymph nodes.[4] In 85% of cases, patients with Rosai-Dorfman disease are in good general health without significant symptoms of the disease.[5] Treatment depends upon the

patient individually and is planned after thorough testing to determine the extent of disease.[6] Ideal treatment, however, has not been established, and there is no ongoing clinical trial.[7]

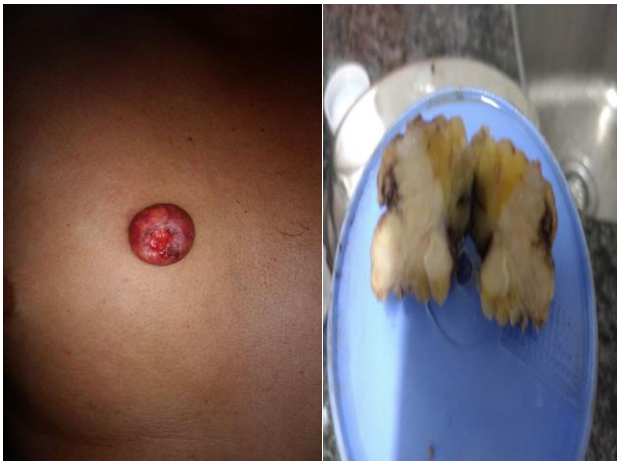

Figure 1: Shows chest lesion clinically and gross of cutsection after surgery shows whitish firm tumor below the epidermis.

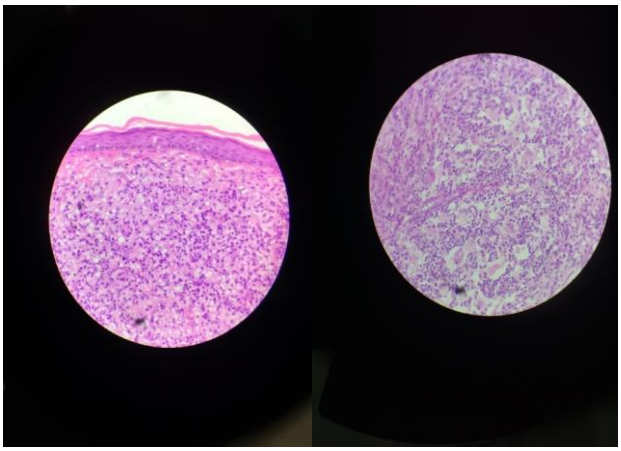

Figure 2: Shows microscopy showing chronic inflammatory cell infiltrate with lymphocytes, plasma cells and histiocytes containing lymphocytes in their pink cytoplasm and demonstrating emperipolesis.

It is believed that 70%–80% of patients recovered from symptoms without treatment, although they may have alternating episodes of exacerbation and remission of symptoms for a long period of time, some patients with severe or persistent disease or cases with risk of organ dysfunction may require treatment with surgery, steroids, and/or chemotherapy.[8,9] Radiation therapy is rare to be used.In Skin Involvement surgical removal is treatment of choice.[10]

CONCLUSION

Rosai- Dorfman disease is a rare disorder which usually involves only lymph nodes and is generally self-limiting. However, in rare instances, can show extranodal involvement like skin & mimic carcinoma clinically.

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