A Case Report

Rosai Dorfman's Disease

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INTRODUCTION

This fascinating disorder is also known as sinus histiocytosis with massive lymphadenopathy (SHML). The most common presentation is massive painless bilateral lymphadenopathy with fever. The disease has a predilection for blacks. In few cases extranodal areas may be involved. Importance of this disease is its capacity to simulate a malignant process, both clinically and pathologically. It has a benign clinical course.

CASE REPORT

A 28 year old male, labourer by occupation presented in Allied Hospital, Faisalabad with bilateral cervical lymphadenopathy. He gave a history of fever for the last 6 months. The fever was low grade. It was noted off and on with no evening rise. On the basis of lymphadenopathy, fever and a raised ESR he was put on anti tuberculous therapy (ATT) which he took for six months. During this time the fever subsided but the lymphadenopathy progressively increased.

The patient was re- examined by another consultant. On examination there was bilateral cervical lymphadenopathy. These lymph nodes were discrete and mobile and the overlying skin looked normal in appearance (Fig 1). There was no history of cough or sputum production however the patient gives history of post nasal drip off and on. There was no significant pallor, hepatosplenomegaly, any other lymphadenopathy or other positive clinical findings.

The blood complete examination was normal. Chest X-ray was normal while that of the head and neck revealed haziness of the paranasal sinuses. Computerised axial scan (CT) of the neck showed bilateral massive lymphadenopathy of significant size. No bony lesion was seen. Magnetic resonance imaging (MRI) was also done. It showed a soft tissue density mass present in all groups of paranasal sinuses including the nasal cavity. The nasal septa were thinned out and eroded but no frank destruction was noted in these bones.

Because of these findings a cervical lymph node biopsy was done and sent for histopathology. The specimen was received in 10% formalin. The lymph node measured 2.5x1.0x1.0 cm and weighed one gram. The cut surface was grey white and homogenous. The whole specimen was submitted totally in 3 blocks which were stained with hematoxylin and eosin. The histological section showed partially maintained nodal architecture. The predominant finding was of moderate to marked histiocytic proliferation. The sinuses revealed pronounced dilatation and were filled with many foamy histiocytes which had round to oval nuclei (Fig 2). No folding or irregularity of the nuclei was seen. The cytoplasm was abundant and revealed engulfed neutrophils and lymphocytes (emperiopoesis). Mitosis were present. Reed Sternberg, lacunar, popcorn or any other atypical cells suggestive of haemato-lymphoid malignancy were not seen. No granulomas, necrosis or any tumour deposits were seen. Immuno-histochemistry was done and showed that S-100 was positive in the histiocytes. On this basis a diagnosis of sinus histiocytosis with massive lymphadenopathy (SHML) (Rosai Dorfman's disease) was made.

Following this fine needle aspiration cytology (FNAC) from the other lymph nodes was also done and the slides stained with Giemsa stain. These revealed many foamy histeocytes with round to oval nuclei and abundant foamy cytoplasm. Some of these histeocytes showed emperio poesis (Fig 3). These cells were seen lying in a background of mature lymphocytes and plasma cells.

The patient was referred to the oncology department from where he was referred to the medical unit. He was started on prednisolone but the lymphadenopathy increased markedly and the patient started having hypertension. He was then tapered off from prednisolone. The size of the lymph nodes which had increased previously started to reduce after drug withdrawl. The patient only had nasal symptoms after this for which he consulted the ENT specialist. Uptil

now he is not any drugs and takes only symptomatic treatment for the post nasal drip. The size of the lymph nodes is static.

DISCUSSION

The synonyms are sinus histiocytosis with massive lymphadenopathy (SHML) and Destombes – Rosai Dorfman's disease. It is a very rare disorder. Probably less than a thousand cases are reported in literature [1]. The aetiology and pathogenesis of this multisystem disorder are unknown but are thought to represent a reactive histeocytic process to an infective agent rather than a neoplastic or other primary condition [2]. The most likely etiological agent is thought to be HHV-8 but this organism is so commonly present in the lymphoid tissue that its role is doubted [3]. Another etiological agent is stimulation of monocyte/ macrophages via macrophage colony stimulating factor (M-CSF) leading to immune suppressive macrophages [4]. Persistent painless lymphadenopathy due to expansion of sinuses infiltrated with benign histeocytes and plasma cells is the characteristic feature of SHML. [5]

SHML registry was initiated in the department of Pathology at Yale University School of Medicine. Drs Foucar, Rosai and Dorfman reviewed 423 cases of SHML and entered them in a registry with special emphasis on the extranodal manifestation of the disease. Most common extranodal sites involved are eyes, ocular adenexae, head and neck, upper respiratory tract, skin, subcutaneous tissue, bone, skeletal muscles, CNS, GIT, salivary gland, gut, thyroid, breast, liver, kidney, heart, cervix etc [6].

Most cases of SHML occur during first or second decade of life. The present case was diagnosed at the age of 28 years however any age group can be affected [7]. The most commonly involved site is the cervical lymph nodes. Extranodal involvement occurs in 30-40 % of the cases most often in the head and neck [8].

Rarely patients with SHML also have malignant lymphoma usually involving anatomic sites different from those involved by SHML [9]. Other common neoplasms associated with SHML are NHL, myeloma, melanoma, papillary carcinoma thyroid and immature teratoma of the ovary [10,11]. An exceptional case of Rosai Dorfman's disease was seen in a 60 year old Japanese where it was seen arising from the meninges [12]. Another unusual presentation is bilateral periorbital swellings which were painless and progressively increased in size in a 12 year old boy [13]. Agarwal A et al reported a study of seven cases (5 nodal and 2 extranodal) with a detailed follow up of 5 patients. Out of these 5 patients 4 had a stable course while 1 developed histiocytic sarcoma after a gap of 4 years [14].

In addition to emperiopoesis (lymphophagocytosis) reactivity for S 100 and CD 68 and non reactivity for CD 1a immune-staining are characteristic features of this histioproliferative disease [15].

CONCLUSION

SHML is a rare disorder affecting any age group commonly presenting as painless lymphadenopathy. The histiocytes are positive for CD 68 and S100 protein. The course of the disease varies. Mostly the lymph nodes regress on their own, however reports are available in which radiotherapy, chemotherapy or surgical debulking is done and found to be effective treatment.

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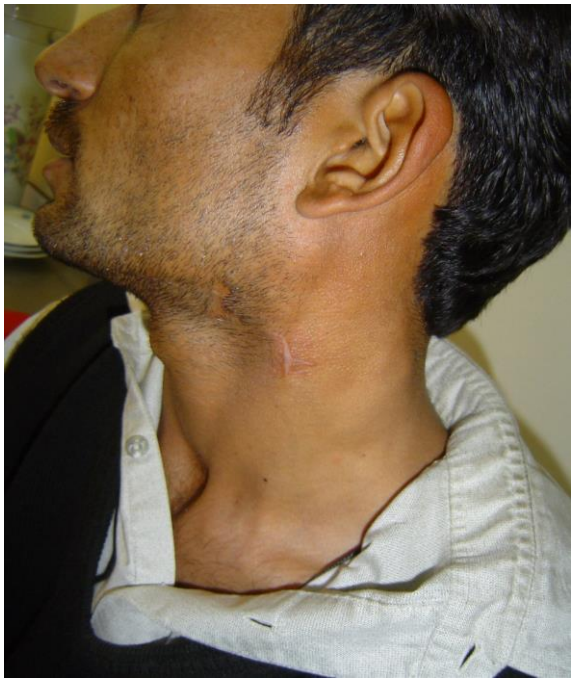

Figure 1
Cervical Lymphadenopathy

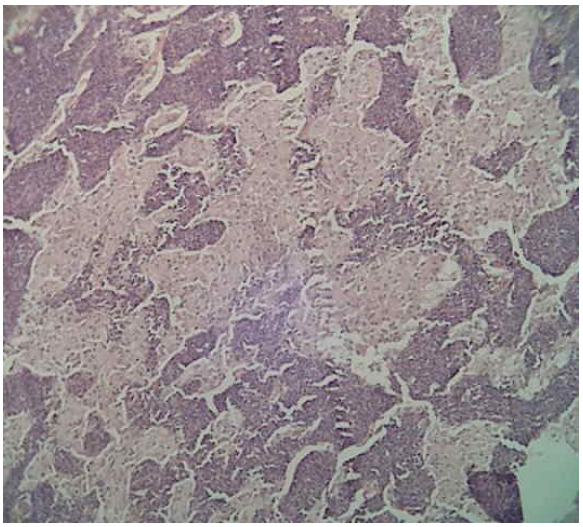

Figure 2
H/E stain showing marked sinus histiocytosis

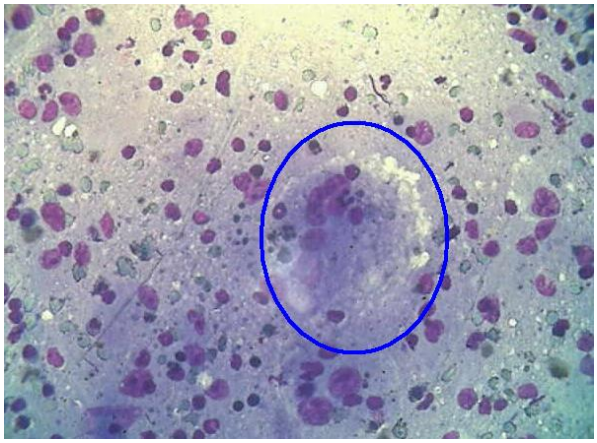

Figure 3
Fine needle aspiration cytology showing Emperiopoesis

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