Benign histocytosis: case report and challenges with its management in a Nigerian Tertiary Hospital

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Abstact

Background: Sinus histocytosis is a rare benign histocytic proliferative disorder affecting several body tissues. Managing histocytosis is associated with a lot of challenges that could result in a delay in initiating therapy. This contributes to poor outcome taking into cognizance the usual problem of late presentation in developing countries.
Methods: We report a case of a 10 year old female child with Sinus histocytosis and outlined the challenges in her management and concomitant intervention we offered.
Results: Findings revealed that there were limitations faced with diagnosing and managing sinus histocytosis. This is a common finding with rare conditions especially in low resource countries.
Conclusion: To improve the quality of care for this group of patients, a multidisciplinary approach to its management is recommended in low resource countries.
Keywords: Sinus histocytosis, lymphadenopathy. Rare, Biopsy,

Introduction

Histocytosis refers to a disorder characterized by accumulation and infiltration of monocytes, macrophages and dendritic cells in affected tissues. Their occurence in the tropics could be said to be rare ostensibly because they are being under-reported. This may be attributed to the challenges with making their diagnosis. They may also be misdiagnosed for other tumours common in the region. Limitations in laboratory facility could be a contributory factor[1]. The incidence of histocytosis is about 4-5 per million of population in western world[2] but in Nigeria only hospital based data are available. Olu-Edo and Egbagbe[3] reported a prevalence of 6.3% among children with generalized lymphadenopathy. The possible aetiological causes includes cellular and immune dysfuction, antecedent viral infections and genetic predisposition[4-6]. These have remained postulations that are yet to be substantiate and therefore the causes of histocytosis remain elusive.

We present a case of a 10 year old Nigerian child with histocytosis with emphasis on the challenges in its management.

Case Report

A 10 year old female presented with multiple chest and facial swellings first noticed a year to presentation. Swellings were first noticed on the chest and later spread to involve the face and neck. Swellings were mostly nodular and not tender. They were not painful and no changes in colouration over affected areas were noticed. No history of fever, cough, convulsion or headache, though there was concomitant weight loss despite good appetite. No history of weakness in her limbs or challenges with ambulating. She is the second and last child of the mother in a seperated family. The mother is a petty trader and she was a primary three pupil who stopped going to school since onset of the illness.

Examination revealed massive facial swelling and peri-orbital oedema (figure 1&2). The mass was nodular, firm with no differential warmth and measured about 14 by 8cm in their widest diameter on the maxillary and mandibular region of the face. She had generalized significant lymphadenopathy ranging between 2 to 3cm (cervical, axillay and occipital) (figure1), Axillary frecklings or cafe-au lait spots was not observed.

There were multiple nodular masses on the anterior chest wall which were more prominent at the peri-clavicular region (figure 2). They were not tender. She also had hepato-splenomegaly.

An initial diagnosis of Rhabdomyosarcoma was made. Other considerations were Lymphoproliferative disorder and Tuberculosis of the skin (Lupus Vulgaris). Initial results of investigation showed leucocytosis (10.9×10^9), eosinophilia, skull X-ray showed an intact lamina dura and no osteolytic bone changes. The bone marrow aspirate showed slightly increased plasma cell with no other abnormal finding. Biopsy specimens were subsequently obtained from different sites of the swelling.

An incisional biopsy from chest swelling revealed a reactive tissue.

Biopsy from the intra-oral extension of swelling showed extensive ulcerated stratified squamous epithelium with focal areas of hyperkeratosis.

A 3rd biopsy from the occipital lymph node showed diffuse sheets of reactive lymphocyte with expansion and distension of sinuses containing numerous histocytes and pigment laden macrophages called “Emperiopoisis”. This was consistent with Sinus Histocytosis. This diagnosis was made 10 weeks into her admission.

She was then commenced on chemotherapy:
Cytosine arabinoside at 1mg/kg/dose 6 hourly for 3 days
Vincristine at 1.5mg/m^2 start on day 1
Prednisolone at 1mg/kg in 4 divided doses for 30 days.

She completed the first course of chemotherapy with no appreciable change in her clinical status. She was scheduled to commence the second course of chemotherapy a fortnight from date of completing the first course, however the mother persistently asked for discharge on account of long hospital stay (3 months) inspite of medical advice against that. She was discharged and could not be contacted subsequently despite several calls and effort to trace her residence.

Identified challenges in the management of our patient are:

1. There was an initial diagnostic challenge because it is a rare disease condition with no data on its prevalence.
2. The parents of our patient were separated and the father did not finance her health bills. Her mother who is a petty trader could not meet up with cost of her treatment.
3. There is no preferred treatment modality for Sinus histocytosis which made it difficult to initiate treatment even following diagnosis.

Figure 1: Lateral view of facial swelling

Figure 2: Massive facial swelling and periorbital oedema

Discussion

Sinus histocytosis is a rare idiopathic benign disorder. Making a diagnosis and managing it could be a herculean task. The challenges faced in resource poor countries towards making diagnosis of rare conditions have been exemplified with our experience. Sinus histocytosis, specifically the Rosai Dorfmann variant has been reported in western literature[7]. A peculiar finding is the deceptive nature of the presentation[7-8]. Misdiagnosis is a common feature with the disease. Rajasekharan *et al*[8] initially misdiagnosed their case as a lymphoma and following multiple biopsies they eventually arrived at the diagnosis.

This may be attributed to the fact that there are no specific clinical features associated with this condition. It has a pan-systemic manifestation and usually classified as nodal and extranodal presentations[9]. Our patient had hepatosplenomegaly but no fever which is common among those with extranodal lesions[9]. Lymph nodes have been reported in 30% of cases and may cause obstruction to neighbouring tissue and organs[10]. Our patient had multiple lymph node swelling but no evidence of obstruction to vital organs was seen. Nodular non tender swellings were prominent findings in our patient. Cutaneous manifestations are said to be the most common extra nodal manifestations[11] and they could present as papules or nodules and usually not tender[12]. Bone involvements are rare probably why it was not seen in this index case.[13] The diagnostic modalities we explored included haematologic assessment, bone marrow examination, fine needle aspiration cytology and histology. Of the four modalities, histology has been acknowledged as the mainstay in histocytosis diagnosis.[14]Immunochemistry may also be helpful, with the positivity of S-100 and lysozyme though this facility was not available in our hospital.[14]

In developing countries the turnover of histology result is hampered by limited resources, poor facilities and the heavy patient load in various facilities[15]. These factors contributed to the prolonged hospital stay and delay in making diagnosis. Another issue we contended with was the various sites we had to take our biopsy tissue from. Initial site was from the chest, which was the primary site of the lesion. A repeat sample was from the intra-oral extension, these initial two sites were however not representative of the lesion. A third sample was from an occipital lymph node, this was representative and gave away the diagnosis. This finding may suggest the superiority of

draining lymph nodes of the lesion over other plausible sites in making diagnosis.

Treatment is not advocated in all situations especially in the absence of constitutional symptoms, spontaneous regression has been reported in 10-20% of cases[16]. Surgery, chemotherapy and radiotherapy are suggested treatment options with variable outcomes[9]. Surgery is indicated for cosmetic reasons and to prevent functional obstruction. Failla *et al*[17] reported superiority of photodynamic therapy over chemotherapy for a resistant scalp lesion. This mode of treatment is not readily available in resource poor countries. Our patient had only one course of chemotherapy, this was a readily available modality of treatment in our domain. As at the time our patient left against medical advice there were no remarkable improvements in the child's lesion though sinus histocytosis is reported to be benign. While on admission there was evidence of social, economic and emotional challenges in the family. Despite the support from the medical team and social workers the mother still preferred the option of discharge against medical advice. Kotwel and Prabhaker have advocated more support for single mothers by both government and non-governmental organizations[18].

Conclusion

Sinus histocytosis remains a rare benign condition. We recommend a multidisciplinary approach to its management to reduce duration of hospital stay and means of facilitating improvement in patient's clinical condition.

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Conflict of interest: Nil