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ACUTE T- CELL LYMPHOBLASTIC LYMPHOMA-A CASE REPORT
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SUMMARY

We highlight the case of a two year old female who presented with a two month history of left posterior auricular swelling. The swelling developed following trauma, was painless and progressively enlarging. After extensive evaluation the mass was noted to be an extramedullary presentation of Acute T cell lymphoblastic lymphoma, which is rare. We decided to present this as a case report because of its unique presentation, diagnostic challenge and we also wish to highlight the importance of immunohistochemistry in diagnosis of malignancies.

INTRODUCTION

Acute T cell lymphoblastic lymphoma is a rare type of non-Hodgins lymphoma that commonly occurs in males, and usually presents as lymphadenopathy. Data on extramedullary presentation of this malignancy is scanty.We hereby present a case of the above in a two year old female seen at the Defense Forces Memorial Hospital in Nairobi, Kenya.

CASE REPORT

B.P two year old female, presented to us with a two month history of left posterior auricular swelling. The swelling developed after trauma, was non tender and progressively enlarging. On examination, she had left post-auricular swelling measuring approximately 5*4cm, non-tender, with associated left upper cervical and occipital lymphadenopathy. Baseline investigations done were as follows: full haemogram-Hb-12.2g/dl,wbc-11.0*10^9/L,lymphocytes-60.7%,plt-484Pg.HIV serology-non reactive.FNAC(fine needle aspirate and cytology) revealed features of an inflammatory swelling possible resolving abscess, or chronic granulomatous inflammation with recommended tissue biopsy.As the tests were being conducted the patient was on analgesics and broad spectrum antibiotics with no response. During the excision biopsy, the surgeon noted an ill-defined mass of the occipitallis muscle extending in all directions, thinnest near the midline and thickest in the post auricular part of the occipitalis muscle. The mass was adherent to the overlying muscle but not to the periosteum. With the above intra-operative findings, the surgeon was of the opinion of a possible malignant lesion, most probably rhabdomyosarcoma.

The patient was then discharged home as she awaited histopathology results. She was then readmitted after one month with the mass having regrown around the same area of excision.

Histology results reported a round blue cell tumour involving the dermis and sub-cutaneous tissue. The same sample was also taken for immunohistochemistry.

Immunohistochemistry results confirmed diagnosis of acute T cell lymphoblastic lymphoma (ATCLL).

Treatment choice of BFM 98 protocol for intermediate risk disease was decided upon and discussed with the parents.

The disease staged using:CT scan abdomen and Chest, but were both normal.CSF cytology was normal. There was no bone marrow involvement based on BMA done.

With the above findings we classified her as Ann Arbor stage 1.

Treatment was initiated as per protocol and was well tolerated. The swelling started to shrink in the pre-induction phase and was completely dissolved by end of induction phase of therapy. She achieved complete remission by end of the first induction. During the course of treatment, she developed CCF(congestive cardiac failure) which was evaluated and attributed to anthracycline-induced myocarditis. It was successfully managed and resolved completely.

At present, she remains stable, now in the long maintenance phase of treatment protocol. She continues to be followed up at the haematology clinic, DFMH.

DISCUSSION

Lymphoblastic lymphoma accounts for about 2% of the non-Hodgkin's lymphomas making them rare.

The T cell sub type comprises 80-90% of lymphoblastic lymphomas and is common in males. This type of lymphoma is common in childhood and teenagers with an average age of 20 years.

The most common site of involvement is the lymph nodes and presentation with lymphadenopathy. However, approximately 50% of the patients may present with dyspnea from a mediastinal mass. Bone marrow involvement is also common at presentation with constitutional symptoms. As seen in this case, our patient presented with an extra nodal disease that is unusual to find in this condition at presentation. Unlike theleukemia/lymphomas seen in this age group, there was no derangement in the full haemogram that would be suggestive of a possible malignancy.

Diagnosis of ATCLL requires excision biopsy evaluation and histology, with confirmation by immunohistochemistry. In our case, Immunohistochemistry was very valuable in defining the specific malignancy that affected the patient.

Staging of the disease is by the Ann Arbor classification although the murphy's staging is used in the paediatric age group. CNS involvement is assessed via a lumbar puncture for cytology which in our case was normal, hence no indication for CNS radiotherapy. Bone marrow evaluation is also necessary for staging and prognostication.

The management for the above is definitive and supportive. The objective is to obtain complete remission on induction therapy. We opted for the BFM 98 protocol intermediate risk for acute leukemia. It was the best evaluated choice for this patient based on published data on efficacy and toxicity profile. However, it is a long protocol timewise and requires maximum understanding and cooperation from parents and relatives to complete treatment without interruption.

Cranial irradiation used in case of CNS involvement.

We present this case report because as noted above, the diagnosis of this condition was challenging and not obvious causing some delay before treatment. Importance of immunohistochemistry is emphasized in diagnosis of malignancies as we have seen in our case. The drawback however, is that this test is not readily available in our part of the world and has significant cost implications to the patient. Further, the role of biopsy and Immunohistochemistry in diagnosis of lymphoid malignancies is clearly demonstrated in our case.

ACKNOWLEDGEMENTS

To the parents of the patient that was paramount in the good response we got with this case. They ensured the patient was available to receive her chemotherapy at the right time. They had patience, were cooperative as we investigated and treated the patient. Much appreciation to the hospital administration that ensured timely availability of the chemotherapeutic agents. The paediatricians, other clinicians, nurses, support staff and all who participated in the successful care are recognised for their timely and valuable contribution.

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