CASE REPORT

Paediatric burkitt lymphoma presenting as a mandible swelling and intussusception

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SUMMARY

Burkitt lymphoma is a rare entity especially in this part of the world. We had an 11-year-old patient presented with swelling of the mandible for a short one-month duration. He was planned for excision biopsy. However developed severe abdominal pain while in the hospital and was diagnosed as intussusception after ultrasound was done. We proceeded with right hemicolectomy and excision of buccal mass. Early recognition and close monitoring of insidious jaw lesions is recommended even in young adults not within the modal age category of endemic Burkitt.

INTRODUCTION

Burkitt lymphoma is a malignant neoplasm of the lymphoreticular system and specifically the B-lymphocytes. It is a form of high grade and aggressive non-Hodgkin B-cell lymphoma. There are three variants or subtypes of Burkitt lymphoma, namely: the endemic (African) type, that has a peak incidence between the ages of five and seven; the sporadic (American) type and the third type that is associated with the acquired immunodeficiency states.[1] The parts of the body affected by this endemic type of tumour are the jaws (40-70%), the abdominal organs (56%), and the central nervous system (30%).[2] In non-endemic regions, the young adults are predominantly affected with the most common presenting feature of abdominal symptoms, while the jaw are affected in only 7-18% of the cases.[3] We hereby present a case of an adolescent with double pathologies of the mandible and abdomen, which has very rarely been reported before.

CASE REPORT

An 11-year-old previously healthy boy presented with left sided facial swelling of one-month duration that increased in size progressively and aggressively. This swelling was associated with difficulty in opening of the mouth leading to reduced oral intake. Physical examination revealed an emaciated, cachexic young boy weighing 17kg. There was a left facial swelling with associated trismus. The swelling measured about 7cm X 7cm, which was hard, fixed, non-tender and smooth in consistency with no skin changes. Intra-oral mass was seen protruding and breaching through the left buccal mucosa. There was no neck swelling palpable and the laryngoscopy examination was normal.

The contrast enhanced computed tomography (CECT) of the neck was performed and it revealed a large soft tissue mass that arises from the medial margin of the left mandible measuring 54mm X 56mm X 64mm (Figure 1a).

The mass was well-circumscribed, mutiloculated with central lucency. It extended up to the temporomandibular joint, inferiorly to the body of left mandible, medially pressing onto the naso-oropharynx and laterally beyond the left mandible. There were features of destructive changes of the left ramus, angle and the body of mandible.

Patient was admitted and planned for excision biopsy. However, on the second day of admission to the ward, patient developed severe abdominal pain for about six hours in duration associated with abdominal tenderness and muscle guarding on abdominal palpation. He also had constipation for the prior three days. Subsequently, we proceeded with ultrasound of the abdomen, which revealed an intussusception. Ultrasound-guided hydrostatic reduction of intussusception was attempted twice but failed.

Surgical intervention was carried out under emergency setting. Intra-operatively it was noted that there was tumour intussusception, terminal ileum intussusception, and ascending colon intussusception. Attempts to reduce the lesion manually failed. Matted lymph nodes were noted at the terminal ileum. Right hemicolectomy was performed. The excisional biopsy of the buccal mass was done at the same time. Histopathology of the left buccal mass and limited right hemicolectomy with the nodes specimen confirmed as Burkitt lymphoma. The tumour cells were diffusely positive for CD20 and CD10. They were negative for CD3, BCL2, MUM1 and TdT. The Ki67 proliferative index was almost 100%. Sheets of fairly monotonous neoplastic lymphoid cells of intermediate size with evenly interspersed reactive tangible body macrophages, resulting in a characteristic "starry-sky" appearance (Figure 1b) was observed. Patient was then referred to a paediatric oncologist for further management. He was started on chemotherapy. He had good progress on follow up. He was tolerating oral food well and had a weight gain of 5kg within six months after completing his chemotherapy regime.

DISCUSSION

Burkitt lymphoma is a subgroup of non-Hodgkin lymphoma with distinct epidemiological, clinical, pathological, immunological and molecular cytogenetic characteristics. The tumour consists of high grade, diffuse, small non-cleaved

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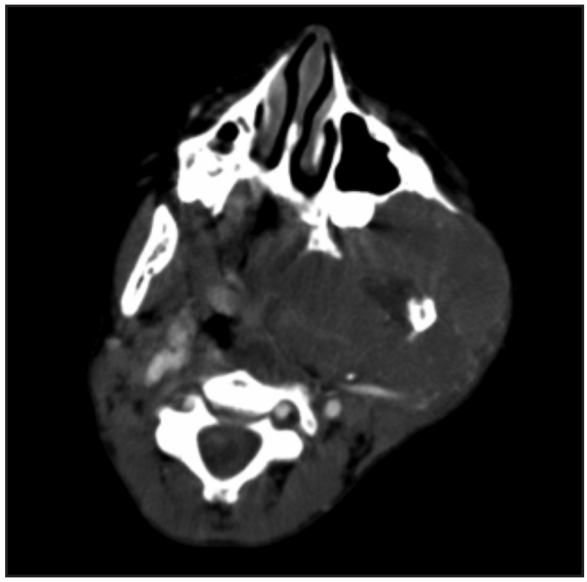

Fig. 1a: CECT neck (axial view) showing the large soft tissue tumour.

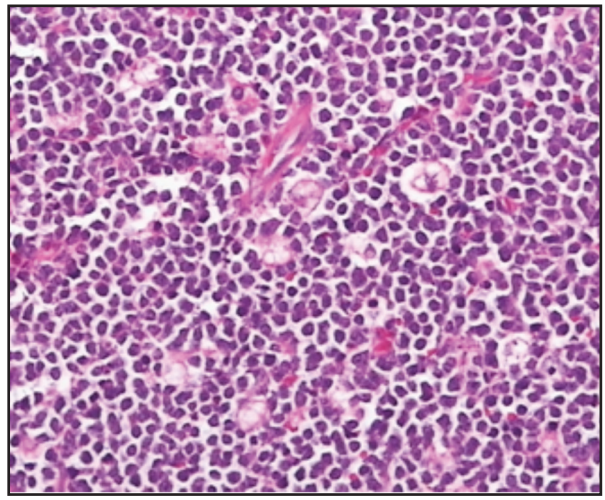

Fig. 1b: Neoplastic intermediate sized lymphoid cells with characteristic "starry-sky" appearance.

B-cell lymphocytes.[4] Head and neck involvement in Burkitt lymphoma includes the facial bones, jaws and other extranodal sites such as intestines. The clinical findings may vary according to the involved anatomical site and the timing of the presentation. Intussusception caused by Burkitt lymphoma, as a cause of acute abdomen, is rare, with symptoms, which are often misleading and make the diagnosis more difficult. The sporadic form regularly presents with abdominal swelling as large mesenteric pelvic or retroperitoneal mass, fullness or pain. In some patients the presentation mimics acute appendicitis.[1] Interestingly, our case recorded that the patient presented with two distant pathological sites at the same time.

The rapidity of volumetric doubling of this neoplasm frequently justifies an acute abdomen presentation that may mimic other less rare diseases.[1] There is strong consideration that EBV as potential etiologic factor of Burkitt lymphoma. The Epstein-Barr virus is as enveloped herpes virus that contains double-strand linear DNA of 70 to 175kb in the nucleocapsid.

In Malaysia, there were only four cases of Burkitt lymphoma reported with jaw and facial presentation. Endemic and sporadic Burkitt lymphoma differ in clinical presentations even though they share similar histological appearance. Both the sporadic and African Burkitt lymphoma involve the abdominal viscera (50-60%). However, in the sporadic Burkitt lymphoma, the jaw is rarely involved.[5] Molecular study was not done for this case. However, in future molecular testing may be of great value in such cases.

CONCLUSION

Early recognition and close monitoring of insidious jaw lesions is recommended even in young adults not within the modal age category of endemic Burkitt. This requires for the clinicians to possess a high clinical index of suspicion. The important role of surgical biopsy for its diagnosis has been amply demonstrated in this report. The implication of this is that prompt diagnosis enhances early commencement of appropriate chemotherapy, which in turn facilitates a better treatment outcome.

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