Primary large B-cell lymphoma of the Ampulla of Vater

Primary lymphoma of the Ampulla of Vater is rare. The clinico-pathological and interesting endoscopic and radiological features of a patient with this disorder is presented.

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Lymphoma of the ampulla of Vater is rare. In a review of 40 cases of cancer involving duodenum or the ampulla of Vater from 1945 to 1970, there were 26 cases of tumors of the ampulla of Vater, but all were carcinoma and none was lymphoma.[1] Amongst primary lymphoma of the ampulla of Vater, the most common histological subtype is follicular lymphoma, followed by marginal zone B-cell lymphoma.[2] However, follicular lymphoma is primarily a nodal disease and primary follicular lymphoma of the gastrointestinal (GI) tract is rare.[3] On the other hand, in a study of 371 Caucasian patients, primary MALT lymphoma of the small bowel accounts for 3.1% of cases.[4] Therefore, the predominance of the follicular and MALT histology in primary lymphoma of the Ampulla of Vater is interesting. On the other hand, while MALT lymphoma of the GI tract is common, stomach is the primary site in the majority of cases.[5]

Case Report

A 73-year-old man presented with hemetemesis. Apart from hemetemesis, there was no fever, night sweat or weight loss. Past medical history included chronic obstructive airway disease, hypertension and cerebrovascular accident. He has been repeatedly admitted because of exacerbations of chronic obstructive airway disease, necessitating intensive care in one admission. Physical examination showed raised jugular venous pressure, parasternal heave and ankle edema, suggestive of pulmonary hypertension. Upper endoscopy and subsequently duodenoscopy using a side-viewing endoscope showed normal mucusa up to the first part of duodenum, swollen mucosa due to lymphoma infiltration at the second part of duodenum (Figure 1, arrows) and a bleeding malignant ulcer measuring 2 centimeters (cm) over the ampulla of Vater. Biopsy showed fragments of tissue extensively infiltrated exclusively by large-sized abnormal lymphoid cells with hyperchromatic nuclei. (Figure 2A; H&E X400) Immunohistochemical study showed that the tumor cells were reactive for CD20, bcl 6 but negative for CD10 and bcl 2. Moreover, 70% of the tumour cells were positive for MIB1. Extensive review of the biopsy did not reveal any low-grade component (Figure 2B; ABC×400), and findings were thus consistent with the diagnosis of diffuse large B cell lymphoma. Contrast enhanced axial CT scans through the upper abdomen showed dilated intrahepatic ducts (Figure 3A; arrowed). Inferiorly at the head of pancreas, a 3.8×2.7 cm heterogenous, contrast enhancing mass at the medial wall of the 2nd part of duodenum in the region of the Ampulla of Vater, (Figure 3B) which extended into the distal common bile duct. (Figure 3B, arrows). It is also

Figure 1.

Figure 2.

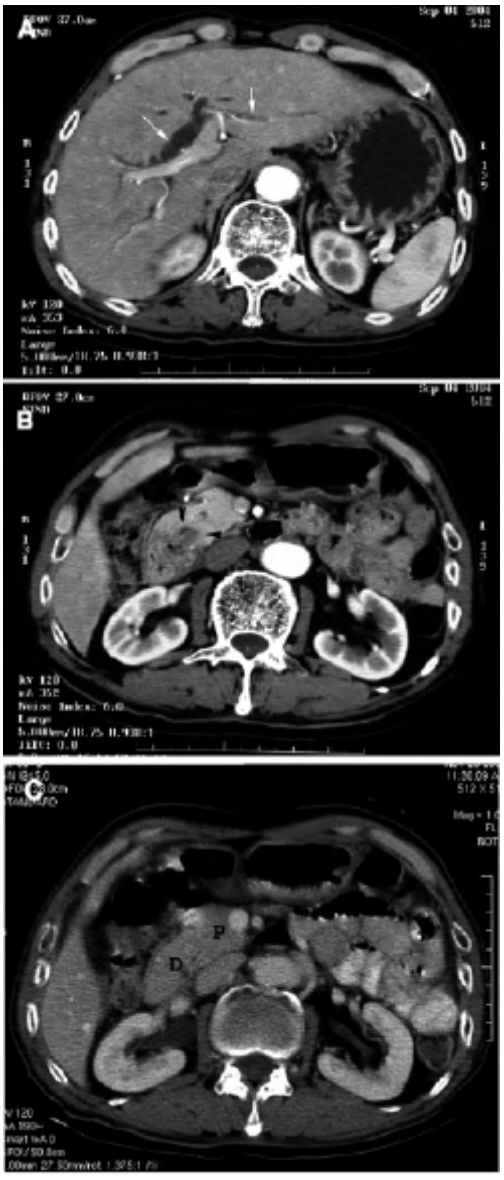

Figure 3.

associated with left para-aortic lymphadenopathy of 1.4 cm. Serum lactate dehydrogenase measure 253 unit/L (normal < 400 U/L) Serum bilirubin measured 27 μmol/L (normal < 19 μmol/L) and alkaline phosphatase measured 188 unit/L (normal < 140U/L). Bilateral trephine showed absence of lymphoma infiltration. Therefore, he had stage II1 primary extranodal lymphoma of the Ampulla of Vater.[6] Echocardiogram showed concentric hypertrophy of the left ventricle with an ejection fraction of 45%. In view of his co-morbid illness and cardiopulmonary status, a non-anthracycline chemotherapy regimen was adopted. He achieved a complete response after three courses of anti-CD20 antibody plus COPP (cyclophosphamide, vincristine, prednisolone and procarbazine) with complete remission. Post treatment scan shows complete resolution of the mass of the tumour at the Ampulla of Vater (Figure 3C) and the common bile duct within the head of pancreas (P) is not dilated (Figure 3C). He was subsequently admitted because of severe exacerbation of chronic obstructive airway disease requiring intubation, and died of aspiration pneumonia.

Discussion

In patients with primary lymphoma of the Ampulla of Vater, presenting symptoms range from upper gastrointestinal bleeding, as in our case, to jaundice, dyspepsia, and symptoms unrelated to the tumor. Some patients may also be complete asymptomatic.[7] For instance primary lymphoma of the Ampulla of Vater in Japanese patients have been diagnosed either from upper endoscopies performed for irritable bowel syndrome or during regular body-check-ups.[7]

In view of the poor cardiorespiratory status in our patient, a non-anthracycline containing regimen was used. However, it is notable that a reduced dose anthracycline-containing regimen can be safely used even in elderly patients older than 80 years.[8] When used as an adjuvant to CHOP (cyclophosphamide, adriamycin, vincristine and prednisolone), rituximab (a chimeric monoclonal anti-CD20 antibody) has been demonstrated to result in higher complete remission rate and superior survival in elderly patients with diffuse large B-cell lymphoma.[9] Therefore, anti-CD20 antibody is used as an adjuvant in this patient but in view of his age and cardiopulmonary status in association with chronic obstructive airway disease, a regimen without anthracycline was used. However, radiotherapy is a viable option, and

involved field radiotherapy has been used successfully in the treatment of MALT lymphoma of the Ampulla of Vater.[10]

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