ipsilateral absence of tonsil and microtia with ectopic salivary gland was reported by Sinha & Singh in 1978. This patient presented with left external ear deformity with conductive hearing loss. His oropharyngeal examination found ipsilateral absence of tonsil and ectopic salivary gland in the posterior part of the tongue. In 1985 Grewal, Hiranandini and Kalgutkar reported 4 cases of congenital external ear deformity with conductive hearing loss along with unnoticed ipsilateral congenital absence of palatine tonsil. In reported cases lowest age was 6 and highest was 18. In our case 28 year old male presented with the features of acute tonsillitis of right tonsil with absence of left tonsil without external ear deformity, hearing loss or any other congenital abnormality. Here absence of tonsil is an incidental finding.

A case presents with congenital external ear deformity one must search for absence of palatine tonsil. But the patient presents with absence of one palatine tonsil without external ear deformity, the findings may remain unnoticed and lately presented following an acute attack of the opposite one.

Congenital unilateral absence of tonsil is a very rare condition. Early diagnosis is possible along with external ear deformity. Agenesis of one tonsil without external ear deformity may have delayed or incidental presentation.

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Back Pain: An Unusual Presentation of Castleman’s Diseases

A 17-year-old school boy from Comilla attended us on 4 August, 2010 with severe episodic back pain, abdominal discomfort and intra-abdominal masses for 10 months. The pain usually appeared once in a week or two, spreading from mid back upwards, gradually deteriorating over months. The first physician he encountered found several intra-abdominal masses for which he underwent abdominal surgery in January, 2010. Two months after surgery his symptoms reappeared with localized abdominal swelling right to the umbilicus, and intermittent high grade fever, drenching night sweats, generalized weakness, anorexia and weight loss. His investigation reports over this period revealed mild normocytic normochromic anaemia and raised ESR. WBC, platelet count, bilirubin, creatinine, LDH, blood urea, ALT all were within normal limit. On 6 January, 2010 he underwent abdominal surgery for lymph node biopsy and histopathological examination of which revealed angiofollicular lymph node hyperplasia, i.e.; Castleman’s disease (CD). On 2 Aug 2010 MRI of lumbosacral spine was advised by neurosurgeon due to persistent back pain which revealed insignificant finding.

Patient attended Department of Haematology in BSMMU after 7 months of diagnosis of Castleman’s disease. On physical examination multiple intra-abdominal masses were found in umbilical region and right iliac fossa, the largest one measuring about 5cm X 5cm, fixed, tender, with smooth surface. Repeat excisional biopsy was important because multicentric CD has potential to evolve into NHL. Previous paraffin impregnated tissue blocks were collected and sent for second opinion. The comment was similar as before, consistent with plasma cell variant (PCV) of CD.

Discussion

Castleman’s disease was first recognized in the 1920s and was further described as a clinicopathologic entity in 1954. In 1956, Dr. Benjamin Castleman and coworkers described a patient with a localized mediastinal mass that on biopsy showed hyperplasia resembling Hassall’s corpuscles of the thymus, as well as capillary proliferation and hyalinization. Histologically, Castleman’s disease is an atypical lymphoproliferative disorder not clearly identified as reactive or neoplastic. While not officially considered a cancer, the overgrowth of lymphatic cells with this disease is similar to lymphoma. The
major difference between CD and lymphoma is the conlatity and loss of architecture in the latter.

Clinically, most CD are unicentric (UCD) and so clinically benign without B symptoms. On the other hand, multicentric CD (MCD) might be clinically indistinguishable from lymphoma. So far, CD has been mostly found in second to seventh decade of life and young patients mostly present with UCD; but our case is clinically MCD in a teenager which is unusual.

Another unusual presentation is the back pain. Pain from retroperitoneal structures may refer to back (e.g., pain from pancreas or kidney), but intra-abdominal lymph node enlargement causing back pain is quite unusual, that compelled neurosurgeons to go for MRI of lumbosacral spine.

Morphologically CD is classified into hyaline vascular variants (HVV) and plasma cell variants (PCV); some authors preferred another class of mixed variants. When combining clinical and morphological features, CD can be classified in 4 major types: unicentric HVV (72%), unicentric PCV (18%), multicentric PCV (10%, like this case) (Figure 1) and rare multicentric HVV (1%)1.

Pathogenesis of UCD is inconclusive, but it is said that IL-6 has a strong role in the pathogenesis of MCD and deletion of IL-6 gene prevents developing Castleman’s disease. It is postulated that HHV-8 produces interleukin 6 and is responsible for lymphoplasmaic proliferation2. That is why anti-IL6 (suramin) and IL-6 receptor antagonist antibody (atlizumab) both are tried for treatment of MCD3.

From the above discussion it is clear that in our case many things should have been done but for unavailability and financial restraint we couldn’t do many tests, namely, PET to see true disease involvement; and HHV-8 PCR and CD20 which guide therapeutic measures where HHV-8 positive cases respond to valganciclovir, and in CD20-positive cases anti-CD20 monoclonal antibody (rituximab) is considered4.

At present, there is no consensus as to the optimal management strategy for MCD. Successful treatment of MCD has been achieved using chemotherapy, with or without prednisone, given at the time of initial diagnosis5. That is why, considering the age and toxicity of chemotherapeutics, he was treated with 4 cycles of ABVD which was completed by December 2010. On follow up for 6 months, there was significant clinical improvement and disappearance of abdominal lymphadenopathy evident by ultrasonography.

Conclusion: Though rare, Castleman’s disease should be considered in patients presenting with single or multiple lymph node enlargement. Due to malignant potential and late complications, patients with multicentric CD should be offered chemotherapy early with adequate explanation and counselling, which might prevent reaching a point of no return.

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References

Constipation – presenting compliant and clinical marker of Parkinson’s disease

To date a large portion of existing literature has reported the high incident of constipation present in Parkinson’s disease patients. Some studies have reported prevalence rates of constipation in Parkinson’s disease (PD) patients as high as 90%6. The disease causes a loss of peristalsis,