Castleman’s disease of the mesocolon: a rare case report

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Abstract — Castleman’s disease is a rare form of localized lymph node hyperplasia of unknown etiology. The sub-types are; hyaline vascular, plasma cell and mixed variant. Clinical subtypes are localized (unicentric) and multicentric. It is reported in all age groups regardless of gender. Hyaline vascular type, accounts for 90% of all cases, often develops in the neck, mediastinum and pulmonary hilum. Its occurrence in the peritoneal cavity is very rare. We present a case in mesocolon of hyaline type in a 39 year female.

Keywords — Castleman’s disease, mesocolon, angiofollicular lymph node hyperplasia.

INTRODUCTION

Castleman’s disease (CD) is a rare form of localized lymph node hyperplasia of unknown etiology. The various histopathological sub-types are; hyaline vascular, plasma cell and mixed variant. Clinical subtypes are localized (unicentric) and multicentric (Eszes et al., 2014). Most common type of unicentric CD is hyaline vascular type. Reported in all age groups regardless of gender (Schaefer et al., 2011). It can occur in any part of the body as nodal/extranodal mass. Common sites are mediastinum (65%), Neck (16%), abdomen (12%) and axilla (3%) (Guo et al., 2012). Its occurrence in the peritoneal cavity is very rare (Miyoshi et al., 2013; Papaziogas et al., 2006).

The definitive diagnosis of CD is based on histopathological examination. Hyaline vascular type is characterized by abnormal follicles with shrunken germinal centres consisting of follicular dendritic cells, ingrowing hyalinized blood vessels and the interfollicular hypervasularization (Schaefer et al., 2011). Plasma cell variant shows hyperplastic germinal centres, intact mantle zone infiltrated by mature plasma cells and interfollicular plasmacytosis (Schaefer et al., 2011). Mixed cellularity is a combination of hyaline vascular and plasma cell type. The hyaline vascular type most commonly presents as a single unicentric mass having a favourable prognosis, whereas the plasma cell type is almost always multicentric and harbours a worse prognosis (Dei-Adomakoh et al., 2013).

CASE REPORT

A 39 year old female presented with abdominal pain since 2 months. Pain was continuous, mild to moderate, and present in epigastric region. No history of altered bowel habits. General physical examination showed the patient was moderately built and well nourished. Laboratory investigations were within normal limits. Family history was insignificant. CT abdomen diagnosis was benign mesenchymal tumour/desmoid tumour/GIST. CT guided FNAC was done which revealed moderate cell yield consisting of benign spindle cells in cohesive sheets. A diagnosis of spindle cell tumour was given. Laprotomy was done, lesion was excised and sent for histopathological examination.

Grossly specimen was irregular, nodular, grey brown mass measuring 8x7x5cms (Figure 1). Cut surface was gritty, grey white to grey brown with focal calcified areas. Microscopy showed structure of lymph node with effacement of normal architecture consisting of small lymphoid follicles with diminished germinal centre surrounded concentrically by mature lymphocytes. The centre of lymphoid follicles showed...
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hyalinised blood vessels along with prominent dendritic cells (Figure 2, 3). Focal areas showed prominent dilated sinusoids, marked fibrosis and calcification. Immunohistochemistry showed CD15, CD30 negative. CD45, CD20, CD3 and CD5 showed diffuse positivity (Figures 4, 5, 6, 7). Finally diagnosis of Hyaline vascular type of Castleman's disease (CD) was offered.

Discussion

Castleman's disease is a rare form of localized lymph node hyperplasia of unknown etiology, also designated as angiofollicular lymph node hyperplasia, giant lymphnode hyperplasia, angiomatous lymphoid hamartoma, benign giant lymphoma (Ioachim H L, 2009). Studies have shown that CD is associated with increased production of cytokine interleukin-6 (Tazi et al., 2011).

Figure 1. Gross photograph of the excised specimen which is irregular and nodular.

Table 1. Shows the clinico-pathological features of various case reports.

| Serial No | Article                        | Age  | Gender | Site                | Histological Variant | IHC Markers (+)                          | Follow up | Prognosis                                                                 |
|-----------|--------------------------------|------|--------|---------------------|----------------------|------------------------------------------|-----------|---------------------------------------------------------------------------|
| 1         | Papaziogas et al. (2006)       | 22yrs| F      | Mesentry           | Hyaline variant      | Plasma cells positive for K & lambda chains | 10mths    | Ten months after operation the patient was free of signs of recurrence   |
| 2         | Schaefer et al. (2011)         | 37yrs| F      | Lower extremity    | Mixed cellularity    | CD23, CD138, KiM4P                       | 3mths     | No evidence of relapse/ma                                               |
| 3         | Tazi et al. (2011)             | 45yrs| M      | Cervical lymph node| Hyaline vascular     | -                                        | 2yrs      | No relapse of CD, but nephrotic syndrome is in remission                |
| 4         | Guo et al. (2012)              | 43yrs| M      | Intraabdominal (pancreatic head) | Hyaline vascular     | -                                        | 1mth      | Tumour markers were reexamined 1 mth after the operation, and CA-19-9 in both patients were within normal range |
|           |                                |      |        |                     |                      |                                          |           |                                                                           |
| 5         | Miyoshi et al. (2013)          | 70yrs| F      | Liver              | Hyaline vascular     | -                                        | Under follow up | No signs of recurrence                                                  |
| 6         | Dei-Adomakoh et al. (2013)     | 55yrs| M      | Axillary lymph node| Plasma cell variant  | CD138, kappa, lambda, CD13, CD3, CD20, Bcl-6, CD23, CD21, IgA, Bcl-2, Bcl-1 | 12mths    | Patient was very well after 6 cycles of chemotherapy                      |
| 7         | Present study (2014)           | 39yrs| F      | Mesocolon          | Hyaline vascular     | CD45, CD20, CD3, CD5                      | Under follow up | No evidence of recurrence till now                                      |

Mths: month; yrs: years; M: male; F: female
It was first described in 1954 by Benjamin Castleman. Gaba et al described the first case along with histopathology in multiple lymph nodes in 1974 (Fajgenbaum et al., 2014). Rachna M et al have reported that CD affects less than 2,00,000 people in the US population (Madan et al., 2012). CD has been reported in all age groups regardless of gender. It can occur in any part of the body as nodal/extranodal mass with predilection for the mediastinum (65%), neck (16%), abdomen (12%) and axilla (3%). Its occurrence in the peritoneal cavity is very rare (Miyoshi et al., 2013; Papaziogas et al., 2006). The present case was in a 39 years female in the mesocolon.

The clinical presentation depends on the clinical type; unicentric/multicentric. Unicentric CD is localized to particular region and is discovered incidentally or due to symptoms related to compression by the mass. Various studies have shown different locations which includes head and neck, intra-bronchial, intra-abdominal (pancreatic, liver, mesocolon), axillary lymph node, cervical lymph node and lower extremity (Dei-Adomakoh et al., 2013; Eszes et al., 2014; Guo et al., 2012; Miyoshi et al., 2013; Papaziogas et al., 2006; Schaefer et al., 2011; Tazi et al., 2011). Table 1 shows the clinic-pathological characteristics of different subtypes of CD as described in the previous studies and the present case.

Most common histological type of unicentric CD is hyaline vascular type. On the other hand plasma cell type presents with signs of chronic inflammation such as fever, arthralgia, raised ESR and weight loss. Multicentric CD presents with multiple lesions and also in association with Kaposi sarcoma, autoimmune disorders and POEMS syndrome (peripheral organomegaly, endocrinopathy; monoclonal gammopathy (M-protein) and skin changes) (Dei-Adomakoh et al., 2013). In present case the lesion was unicentric and histologically hyaline vascular type.

The definitive diagnosis of CD is based on histopathological examination. The histopathological subtypes of CD are hyaline vascular, plasma cell and mixed variant. On microscopic examination the hyaline vascular type is characterized by lymphoid follicles with shrunken germinal centres consisting of follicular dendritic cells, ingrowing hyalinized blood vessels and the interfollicular hypervasularization (Schaefer et al., 2011).

Plasma cell variant shows hyperplastic germinal centers, intact mantle zone infiltrated by mature plasma cells and interfollicular plasmacytosis. Mixed cellularity is a combination of hyaline vascular and plasma cell type (Schaefer et al., 2011).
The differential diagnoses of CD are other lymphoproliferative disorders especially lymphomas with plasmablastic features. HIV, HHV8 and EBV are associated with both plasmablastic lymphomas and CD. “B” cells are known to be characterized by CD5 expression in the expanded mantle zones of CD. CD shows positivity for CD45, CD20, CD3 and CD5 as in our case.

For localized unicentric variant, surgical resection of the affected lymph node is curative. Whereas MCD requires aggressive systemic therapy (Dei-Adomakoh et al., 2013). The hyaline vascular type most commonly presents a single unicentric mass having a favourable prognosis, whereas the plasma cell type is almost always multicentric and has a worse prognosis. The present case was followed for one year which was uneventful.

**CONCLUSION**

To conclude, one should be aware of CD, its clinical/histological types, prognosis for its early diagnosis and timely treatment.

**Abbreviations**

CD: Castleman’s disease, FNAC: Fine needle aspiration cytology, CT: Computerised tomography, GIST: Gastrointestinal stromal tumour, ESR: Erythrocyte sedimentation rate, MCD: Multicentric Castleman’s disease.

**Competing interests**

The authors declare that they have no competing interests.

**Authors’ contributions**

Mythri MB – Data collection, writing script, photography. Kalyani R – concept, data collection, revision of manuscript, editing manuscript, selecting gross and microphotograph images. Srinivas Murthy V – editing the manuscript.

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