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Abstract- Castleman's disease is a rare lymphoproliferative disease, that can either be unicentric or multicentric in presentation. It can affect various sites such as chest, neck, abdomen, and many others. Unicentric Castleman's disease commonly affects the mediastinum, arising in the anterior and middle compartment and less commonly the posterior compartment. Patients affected with the disease are usually asymptomatic, however, they may present with non-specific symptoms such as cough and dyspnea. The treatment of Unicetric Castleman's disease is surgical resection. In this case report we present a case of posterior mediastinal unicentric Castleman's tumor, which was completely resected via robotic surgery.

Keywords: *castleman's disease, posterior mediastinal mass, robotic resection.*

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Robotic Resection of a Posterior Mediastinal Unicentric Castleman's Tumor

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Abstract- **Castleman's disease is a rare lymphoproliferative disease, that can either be unicentric or multicentric in presentation. It can affect various sites such as chest, neck, abdomen, and many others. Unicentric Castleman's disease commonly affects the mediastinum, arising in the anterior and middle compartment and less commonly the posterior compartment. Patients affected with the disease are usually asymptomatic, however, they may present with non-specific symptoms such as cough and dyspnea. The treatment of Unicetric Castleman's disease is surgical resection. In this case report we present a case of posterior mediastinal unicentric Castleman's tumor, which was completely resected via robotic surgery.**

Keywords: castleman's disease, posterior mediastinal mass, robotic resection.

I. Introduction

Castleman's disease (CD) is an uncommon lymphoproliferative disorder, of unknown etiology that was first reported by Castleman and Towne in 1954 [1]. Castleman's disease can be classified into 2 main categories, unicentric (localized) and multicentric (systemic) [4]. Castlemna's disease is often asymptomatic. The unicentric type of Castleman's disease is treated by complete surgical resection of the lesion and no further treatment is required after the surgery. In this paper we report a first case of a 43 year old female, presented with constitutional symptoms and a mediastinal mass that was treated by complete surgical resection utilizing Davinci Si robotic system.

II. Case Report

A 43 year old female patient, previously healthy, presented with 20 days history of persistent fever associated with rigors, weight loss, generalized body ache and fatigue. General physical examination was unremarkable. Routine lab tests revealed a microcytic anemia with a hemoglobin level 10.2 g/dL (N: 12-16 g/dL), MCV 73.8 fL (N: 82-97 fL), MCH 23.7 (N:27-33 pg); the rest of the lab tests were within normal limit. Pan CT done and it showed left posterior mediastinal retrocrural paravertebreal, well defined oval solid mass lesion measuring about 54 x 38 x 50 mm, with surrounding retrocrural lymph nodes sharing the same capsule largest suggestive of mediastinal neurogenic tumor likely paraganglioma. MRI of the chest done and it showed well defined paravertebral soft tissue lesion extending from D5 to D12 on the left side measuring 5 x 5 x 5 cm (fig 1, 2). The patient underwent complete excision of the lesion via a minimally invasive procedure with Robotic assisted thoracic surgery (Da Vinci Si model) 3 ports were used. The excised tissue was sent for histopathological testing.

a) Pathologic Findings

The specimen is received fixed in 10% buffered formalin, labeled with patient ID and as "Posterior mediastinal mass". Gross assessment of the specimen revealed a brown firm mass with nodular surface measuring 6.0 x 4.5 x 3.5 cm. Cut surface is brown in color with scattered tan tiny nodules. Another specimen was also received fixed, and labeled as "Pulmonary ligament lymph node". It consisted of a brown nodular tissue measuring 1.2x0.7x0.7cm. Cut surface showed similar changes to the large mass; other small lymph nodes were also included. Microscopic assessment of the mediastinal mass showed an enlarged lymph node composed of atretic small follicles with expanded intervening interfollicular region. Hyalinized vessels traversing the follicles are evident – lollipop sign. The follicular center is replaced by dendrocytes without evidence of atypia. The follicles are surrounded by thickened mantle zone in the form of concentric layering of lymphocytes- which is called as onion skinning. The interfollicular zone is replaced by hyperplastic, some hyalinized, venules and infiltrated by mature lymphocytes. The lymph node septa are thickened and sclerotic. Surrounding the mass, there was a vasculo-adipose tissue containing few peripheral nerves (fig 3). There is no evidence of associated lymphoma. No thymic tissue identified. The other small lymph nodes included show reactive changes only. The findings are of Castleman's disease, hyaline vascular type.

Post-operative follow up the patient had uneventful recovery with fever subsiding.

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The follicles are surrounded by concentric layering of lymphocytes- onion sin appearance- and the follicular center is replaced by dendrocytes. Hyalinized vessels are seen traversing the follicles (lollipop sign).

b) Comment

Castleman's disease (CD) is an uncommon disease of unknown etiology that is characterized by angiofollicular lymph node hyperplasia. There are 2 clinical forms of the disease: localized (unicentric) and systemic (multicentric) form. Histologically the disease can be classified into hyaline vascular type, plasma cell type and mixed type. In most case, the lesion were characterized by prominent vascular proliferation and hyalinzation [3]. In the case we presented her it was of a hyaline vascular type.

Clinical features of CD varies depending of the form of the disease. People with unicentric Castleman's disease (UCD) usually present with a slow growing mass in asymptomatic patients with no gender or race predominance [4]. Common sites of presentation in UCD include the chest (30%), neck (23%), abdomen (20%), retroperitoneum (17%), and, rarely, the axilla (5%), groin (3%), or pelvis (2%) [7]. The disease commonly affect the mediastinum; usually involving the anterior and middle compartment, but rarely the posterior compartment. Only 9 cases of posterior unicentric castleman's disease were reported in the literature. [5, 6].

Patients with intrathoracic disease involvement may present with cough, dyspnea, hemoptysis or chest discomfort, where as abdominal, retroperitoneal, and pelvic disease may present with abdominal or back discomfort [2]. On the other hand, Patients with multicentric Castleman's disease (MCD) usually present with systemic inflammatory manifestations like fever, night sweats, weight loss and fatigue. The physical examination in these patients usually reveals generalized lymphadenopathy, hepatosplenomegaly and signs of fluid retention. Common hematological abnormalities include anemia, elevated inflammatory markers, hypergammaglobulinemia, and hypoalbuminemia. [8] In this paper, we reported a case of UCD that presented with systemic symptoms including fever, weight loss, and fatigue, which is usually a presentation of a MCD.

Histologically the disease can be classified into hyaline vascular type, plasma cell type and mixed type. In most case, the lesions were characterized by hyaline vascular type, which is characterized by increased numbers of small, hyalinized blood vessels within and between follicles with obliteration of the medullary sinuses. [3] The case we presented above was of a hyaline vascular type.

The management of CD depends of the type of the disease. The management of UCD is complete surgical resection; in case the mass is not amenable for resection or the patient is not candidate for surgery, radiotherapy can be used. All 9 cases of posterior mediastinal CD that were reported were surgically resected via open surgical resection, either thoracotomy or sternotomy [5,6].This case is the first posterior mediastinal CD to be completely resected Utilizing robotic surgery (DaVinci SI).

On the other hand, treatment of MCD is include Rituximab, antiretroviral therapy, glucocorticoids, cytotoxic chemotherapy, and anti-Interleukin 6 Therapy. Rituximab can be used as monotherapy for patients with MCD. If the patient with MCD is HIV positive antiretroviral therapy showed be added. Glucocorticoids offer a short term control of symptoms and are usually helpful as an initial adjunct for acutely symptomatic disease. Chemotherapy can be used in cases of relapses or refractory MCD. [8]

III. Conclusion

Unicentric Castleman's disease is rare, and rarely found as posterior mediastinal mass. It may present with systemic manifestation. The treatment of choice is complete surgical resection ,in this case it was demonstrated that surgical resection can be performed using robotic assisted surgery .

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