Harridas M[1], Yusuf S[2], Kori AN[3], Abd Rahman A[4], Jalani N[4], Mohamad I[1]

[1]Department of Otorhinolaryngology-Head & Neck Surgery, School of Medical Sciences, Universiti Sains Malaysia, Health Campus, 16150 Kota Bharu, Kelantan, Malaysia

[2]Department of Otorhinolaryngology, Hospital Tengku Ampuan Afzan, 25100 Kuantan, Pahang, Malaysia

[3]Department of Hematology, Hospital Tengku Ampuan Afzan, 25100 Kuantan, Pahang, Malaysia

[4]Department of Pathology, Hospital Tengku Ampuan Afzan, 25100 Kuantan, Pahang, Malaysia

*Corresponding author
Irfan Mohamad
irfankb@usm.my

Received 13th Nov 2021
Revised 20th July 2022
Accepted 11th April 2023
Published Online 11th June 2023

A Rare Case of Castleman Disease of the Neck: Unicentric Type with Plasma Variant

Abstract – Castleman disease (giant or angio follicular lymph node hyperplasia) is an uncommon lymphoproliferative disorder which is localised to single lymph node (unicentric) or multiple lymph node level systemically (multicentric). It is a very rare disorder characterised by non-cancerous growths. The most common sites of this are mainly thorax (mediastinum or lung hilum) and abdomen. It rarely occurs in the head and neck area. We report a 17-year-old girl who had left neck swelling for 5 years. The excised mass was histopathologically confirmed as Castleman of a rare plasma cell variant, treated with adjuvant radiotherapy. Follow-up after one year showed no recurrence.

Keywords – Castleman disease, lymphoproliferative disorder, non-cancerous, plasma cell

1 INTRODUCTION

Castleman disease (CD) is rare lymphoproliferative disorder, which is commonly found in mediastinum and lung hila. Clinically it is divided in two types: unicentric Castleman Disease (UCD) and multicentric Castleman Disease (MCD). UCD is usually asymptomatic and presents as a mass or swelling while a MCD characterized by fever with chills, anaemia, generalized lymphadenopathy, hepatosplenomegaly and with a more aggressive behaviour [1]. UCD usually presents in young adults with localized masses in the mediastinum (60-75%), neck (20%) or less commonly intra-abdominal masses (10%). MCD are less common than the localized variant [2]. Histologically, the disease is also classified into two separate subtypes: the hyaline vascular and plasma cell variants [2].

2 CASE REPORT

A 17-year-old female presented with left neck swelling for 5 years which gradually increased in size and painless. There were no constitutional symptoms like fever, weight loss, night sweats, reduce appetite, or fatigue.

There was no history of pain in the throat, dysphagia, dyspnoea, change of voice or tuberculosis exposure. On examination there was a swelling at the left infra-auricular region extending to left submandibular region measuring (6cmx7cm), which was firm and non-tender (Figure 1).

There was no other lymph node palpable. Nasoendoscopy examination findings were unremarkable. Computed tomography (CT) showed homogenously enhancing left neck lesion compressing onto the adjacent right internal jugular vein (Figures 2). Incisional biopsy showed hyperplastic follicles with prominent germinal centers surrounded by massive infiltration by plasma cells (Figure 3). There was no hyaline vascular proliferation seen. Patient was then scheduled for excision of the left neck mass under general anaesthesia. The post-operative period was uneventful.

Histopathological examination showed the follicles are positive to CD10 and negative to BCL-2, surrounded by sheets of mature plasma cells (Figure 4). There was no evidence of malignancy.

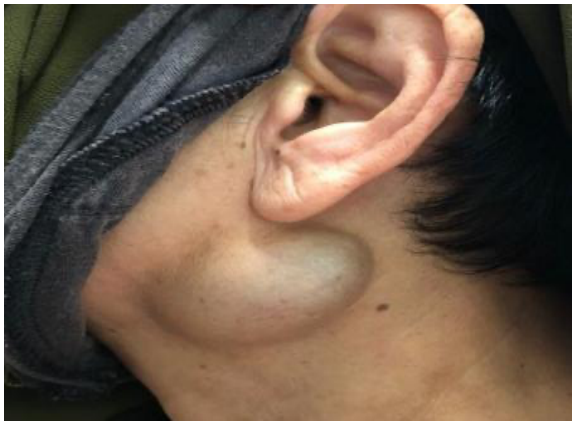

Figure 1. Left submandibular swelling measuring 6 cm x 7 cm

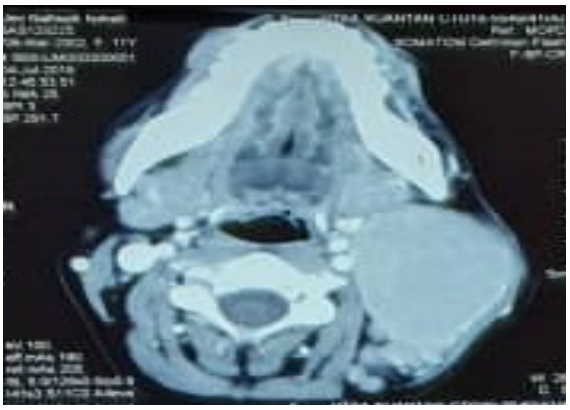

Figure 2A. CT (axial) of the neck shows homogenously enhancing left neck lesion compresses onto the adjacent right internal jugular vein

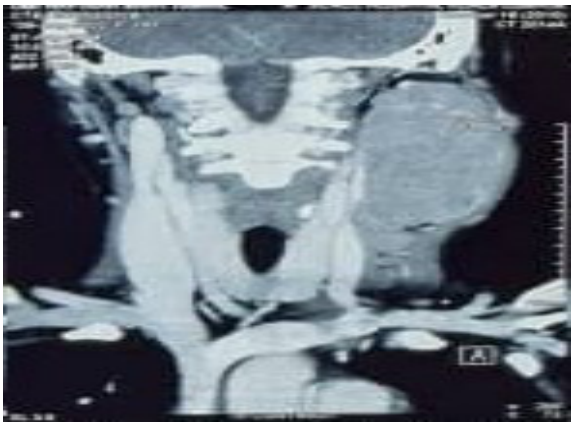

Figure 2B. The similar lesion as seen on CT coronal view

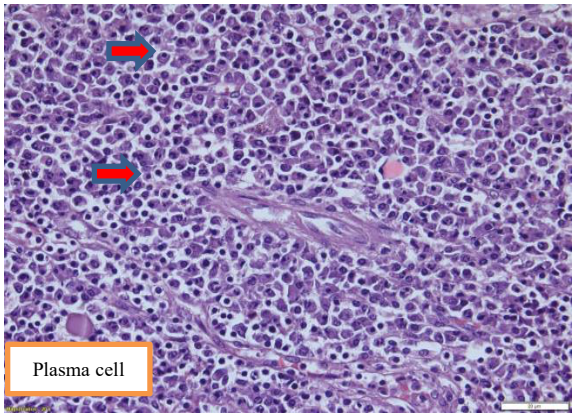

Figure 3. Section shows the plasma cell (original magnification x 20)

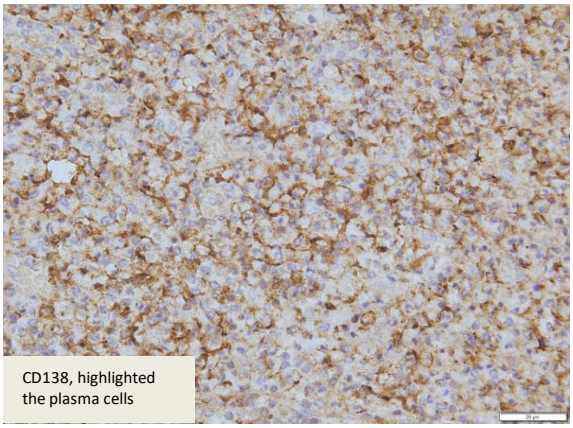

Figure 4A. Section shows plasma cell under CD138 stain (original magnification x 20)

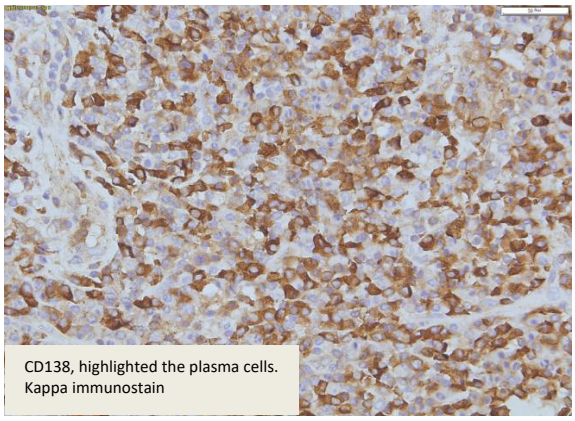

Figure 4B. Section shows plasma cell using Kappa immunostain (original magnification x 20)

Peripheral blood film showed rouleaux formation with occasional lymphoplasmacytoid cells, which suggested plasma cell disorder. Hence, bone marrow aspiration was performed. The bone marrow findings were normocellular marrow with increased bone marrow plasma cells (12%). Bone marrow trephine biopsy result shows increased bone marrow plasma cells with no evidence of clonality seen. Other additional tests such as Serum /urine electrophoresis and B-2 microglobulin were normal.

All other routine lab tests like erythrocyte sedimentation rate, renal profile, liver function test, calcium, lactate dehydrogenase and viral screening were within normal limits. Post-operatively, the patient was well and completed radiotherapy. The patient is on regular follow-up and up to one-year visits, there was no recurrence.

3 DISCUSSION

Castleman Disease was originally described by Dr. Benjamin Castleman in 1954. Three types of CD have been identified. These are the hyaline-vascular (HV) type (about 90% of individuals), plasma cell (PC) type, and mixed variant (MV) type. There are 2 different clinical entities: the unicentric type which only one anatomic lymph node is affected and the multicentric type characterized by generalized lymphadenopathy, constitutional symptoms, and more aggressive clinical course [3].

UCD is the most common type and consists of an isolated benign lymphoproliferative disorder of young adults that is not associated with human herpes virus 8 (HHV8) infection and is usually curable with surgical resection. The vast majority of patients are asymptomatic.

The definitive diagnosis is based on post-operative pathological findings. Once CD is diagnosed, MCD must be ruled out. In addition, UCD may be associated with an increased risk of lymphoma (B-cell non-Hodgkin's lymphoma and Hodgkin lymphoma) [4]. The definitive diagnosis is based on post-operative pathological findings.

This patient is a rare case of UCD with plasmacytic type. Unlike the common UCD, which is of hyaline type, plasmacytic UCD may progress to MCD and is highly associated with HHV8 [4]. To exclude any bone marrow infiltration or lymphoma; blood tests, bone marrow trephine aspiration and biopsy test, imaging and a biopsy of an affected lymph node will be carried out with HHV8 antibodies and viral screening. In UCD, surgical resection of the mass is a standardized and preferred treatment protocol the curative ratio can reach 95%. Surgical treatment can achieve a cure rate of approximately 100%, either with hyaline type or plasma type [5]. In this case of UCD plasmacytic type, surgical resection was carried out combine with radiotherapy. Different treatment options have been described in the literature that usually involves chemotherapy [4], radiotherapy [4], immune modulators [7], or monoclonal antibodies [6].

Plasmacytic type or plasmablastic type may and usually progress to MCD and highly associated with HHV8. It is treatable on relapse with CD20 monoclonal antibodies or anti-IL-6 monoclonal antibodies. The recurrence rate with this treatment is low [4]. In this case, after one-year follow-up, post-surgical resection combined with radiotherapy in UCD plasmacytic type showed no recurrence. Patients with UCD without systemic involvement should have an additional radiological assessment 6-12 months such as a CT scan of the neck after initial therapy, to verify there is no recurrence.

4 CONCLUSION

This case is presented for its rarity in view of unicentric with plasma type variant, with no recurrence and progression to multicentric type, post-surgical and radiotherapy treatment. Cervical lymph nodes are involved by CD and may be confused with other common causes of cervical lymphadenopathy like tuberculosis and nodal secondaries. Surgical removal of the tumors in the Unicentric Plasma type of CD is the treatment of choice combined together with radiotherapy for a better outcome.

REFERENCES

[1] Castleman B. Case records of the Massachusetts General Hospital; Case 40011 (Hyperplasia of mediastinal lymph nodes). New England Journal of Medicine. 1954;250:26-30.

[2] Melkundi RS, Prasad KC, Jalisatgi RR, Swami G, Karunasagar A. Unicentric Castlemans disease: unusual disorder of the neck a case review. Journal of Clinical and Diagnostic Research: JCDR. 2015;9(4):MD03.

[3] Chronowski GM, Ha CS, Wilder RB, Cabanillas F, Manning J, Cox JD. Treatment of unicentric and multicentric Castleman disease and the role of radiotherapy. Cancer: Interdisciplinary International Journal of the American Cancer Society. 2001;92(3):670-6.

[4] Larroche C, Cacoub P, Soulier J, Oksenhendler E, Clauvel JP, Piette JC, Raphael M. Castleman's disease and lymphoma: Report of eight cases in HIV-negative patients and literature review. American Journal of Hematology. 2002;69(2):119-26.

[5] Talat N, Belgaumkar AP, Schulte KM. Surgery in Castleman's disease: a systematic review of 404 published cases. Annals of Surgery. 2012;255(4):677-84.
[6] Beck JT, Hsu SM, Wijdenes J, Bataille R, Klein B, Vesole D, Hayden K, Jagannath S, Barlogie B. Alleviation of systemic manifestations of Castleman's disease by monoclonal anti-interleukin-6 antibody. New England Journal of Medicine. 1994;330(9):602-5.
[7] Jung CP, Emmerich B, Goebel FD, Bogner JR. Successful treatment of a patient with HIV-associated multicentric Castleman disease (MCD) with thalidomide. American Journal of Hematology. 2004;75(3):176-7.