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*Case Report*

# Castleman's Disease in a Kidney Failure Patient Diagnosed Incidentally During Transplantation

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## Abstract

**Introduction**: Castleman's disease is a rare lymphoid disorder. It comprises two pathological entities. These are the hyaline-vascular type which is usually localized (uni-centeric) and the plasma-cell type which is usually multicenteric and rather aggressive.

**Case report**: Here we present a 53 years old Sudanese male who underwent kidney transplantation in August 2009 from a related live-donor.

During transplantation, he was accidently found to have an abnormal looking lymph node at the site of the graft bed. The lymph node was totally excised and sent for histopathology. Based on the histopathological examination, the diagnosis of Castleman's disease of the hyaline vascular type was made. The patient gained normal graft function and was maintained on tacrolimus, azathioprine and prednisolone.

He maintained normal graft function for more than twelve months post transplantation with a serum creatinine level of 0.9 mg/dl. He remained free from recurrence of Castleman's disease during the follow up period.

**Conclusion**: Unicenteric Castleman's disease may be completely asymptomatic. Surgical excision of the lesion was curative for our patient despite maintenance on immunosuppression.

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## Introduction

First described in 1956, Castleman's disease is a rare lymphoid disorder with characteristic lymphoid tissue hyperplasia [1]. It may be occasionally encountered in clinical practice and needs to be recognized and managed [2-5]. It comprises a group of rare lymphoproliferative disorders sharing characteristic clinical and histological features, and may be accompanied by a marked systemic inflammatory response.

Two histological patterns of lymph nodes involvement were described in Castleman's disease: the hyaline-vascular and plasma-cell types. The hyaline-vascular type is more common (80-90%) and tends to be localized in one lymph node and asymptomatic while the plasma cell type has a more aggressive course and is usually multifocal with systemic manifestations [3]. Based on radiological findings, Castleman's disease can also be classified as unicenteric or multicenteric types. Surgery is usually curative for the localized type [4], while the multicenteric type requires drug therapy.

Kidney transplantation is the best available management for patients with end stage renal disease (ESRD) despite potential complications. This paper describes a Sudanese patient with ESRD who was incidentally found to have unicenteric Castleman's disease of an inguinal lymph node.

**Figure 1: Hyaline material in a large follicle (Yellow arrow). The white arrow points at blood vessels in the follicle and inter-follicular zone (White arrow)**

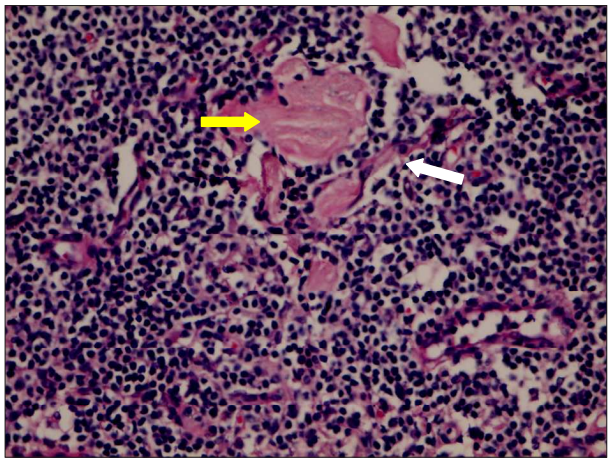
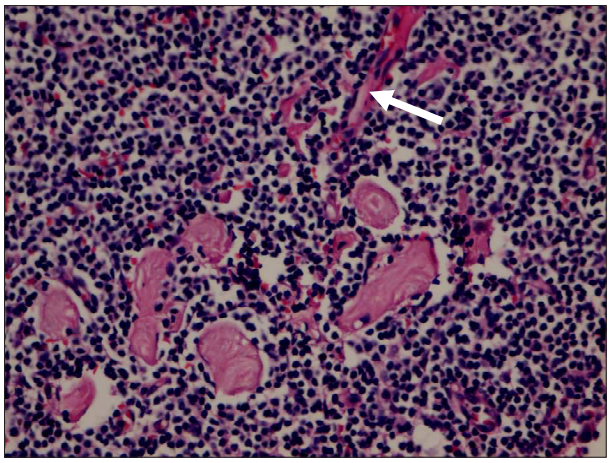


**Figure 2: A small vessel feeding into a follicle containing hyaline material (White arrow)**



## Case report

A fifty three year-old Sudanese male who had uncontrolled hypertension for two years and gout for more than 20 years, was diagnosed to have ESRD in May 2009 and started on regular hemodialysis. He underwent right ureteric dilatation and left ureteric lithotomy in 1981 and appendicectomy in early childhood. He had no history of tuberculosis, diabetes mellitus or lymphoproliferative disease. His family had no history of renal disease but they did have a strong history of hypertension and gout. He does not smoke, abuse drugs or drink alcohol. He presented for kidney transplantation three months after starting dialysis. His clinical examination was unremarkable with a blood pressure of 120/80 mmHg and body mass index of 26 kg/m$^2$. His abdomen was soft, of normal contour with grid-iron scar in the right iliac fossa as well as right and left renal scars. There were neither palpable abdominal organs nor palpable lymph nodes. Sonogrophic examination revealed bilateral small kidneys but no other significant findings. Doppler ultrasound of the pelvic and lower limbs vessels was normal. The patient underwent left iliac fossa renal transplantation in August 2009. During the operation a right inguinal lymph node was found. It was totally excised and sent for histopathological examination.

Examination of several sections showed preserved architecture of the node. Scattered in the follicles and inter-follicular area were many hyaline masses that were negative for amyloid (Figure 1). Some of the hyaline masses contained small blood vessels (Figure 2).There was also a mild degree of sinus cell hyperplasia. The diagnosis of the vascular hyaline type of Castleman's disease was made.

The patient had immediate graft function with no complications, and was maintained on tacrolimus, azathioprine and prednisolone. During more than 12 months of follow up, he continued to have stable graft function with a serum creatinine level of 1 mg/dl. There were no symptoms or sings indicating recurrence of the disease.

## Discussion

Castleman's disease is a rare lymphoproliferative disorder the diagnosis of which may be difficult and challenging. In itself, the localized form may not be a contraindication to transplantation since surgical excision of the lesion is curative for the disease [6].

The multicentric variety is associated with severe systemic manifestations such as hemolysis and involvement of kidneys and the hematopoietic system [3]. At times, it may have an inexorable clinical course. Multicentric disease often necessitates aggressive systemic therapy and portends a poor outcome [6], although recent reports have documented complete remission in response to anti-CD20 monoclonal antibody (rituximab) [5, 7]. The disease is herpesvirus-8-related and may also develop in patients infected with human immunodeficiency virus (HIV) [5]. The patient described herein was tested negative for HIV during pre-transplant screening.

## Conclusion

To our knowledge, this is the first transplanted Sudanese patient diagnosed to have Castleman's disease. Surgical excision of the lesion was curative in our patient.

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