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Retroperitoneal Fibrosis Masquerading as Malignancy: A Clinical Dilemma-Case Report

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Abstract

Retroperitoneal fibrosis is a rare vascular connective tissue disease of unknown aetiology, typically presenting as inflammation within the retroperitoneum with the resultant mass effect of the pathological processes involved compress the organs in the retroperitoneum: Kidneys, ureter, aorta and pancreas. The disease was first described by John Kelso Ormond in 1948 on a patient who presented with bilateral ureteral obstruction secondary to an inflammatory retroperitoneal process [2,3]. Its incidence is at 1.38 cases per 100,000 [1]. It has a male predominance of between 1.6 to 3 fold [1,7]. There have been associations of retroperitoneal fibrosis with various autoimmune conditions.Immunosuppressive treatment modalities have been effective on its control and management thus indirectly inferring to an autoimmune process as a key factor behind its pathophysiology [4,5]. Radiological diagnosis through CT remains a gold standard with biopsy being a controversial option of obtaining diagnosis [5,17-21]. Main stay treatment has been use of glucocorticoids as 1st line, with the disease modifying anti-rheumatic drugs (DMARDS) as a second option where the disease has been found to be steroid resistant [6]. For the scope of this paper, we will describe a case managed at the Karen hospital in Nairobi, Kenya in September 2017.

Keywords: Retroperitoneal Fibrosis; Lymphoma; Retroperitoneal Mass.

Material & Methods

History: 39-year-old Male, known hypertensive since October 2016, presented with elevated blood pressureand occasional palpitations. He reported no history of chest pain, nor difficulty in breathing and lower limb swelling. He wasa teetotaller. On examination, blood pressure was noted to be elevated at 182/130. There were no other abnormal examination findings. ECG recorded a normal sinus rhythm. 2D-ECHO was essentially a normal study.

Laboratory Findings: Patient was evaluated for secondary causes of hypertension and palpitations. Key abnormal laboratory findings were the marginally deranged liver function tests as recorded: Total Bilirubin 18.07umol/l High, Conjugated Bilirubin 10.08umol/l, Indirect Bilirubin 7.99umol/l, Alkaline Phosphatase 315 U/l, AST (SGOT)128U/l0–50 High, Alanine aminotranferase 129U/l0 – 50High, Gamma GT411. Other baseline blood tests done included haemogram, (Hb 15.9, Wbc 6.76, platelets 222), Urea electrolyte, Urine VMA, Cortisol, Thyroids Function Tests, and Parathyroid hormone levels were within normal ranges.

Radiological Findings

With no cause for his hypertension isolated from the laboratory findings, a Renal Doppler ultrasound done showed an increased resistance index of the

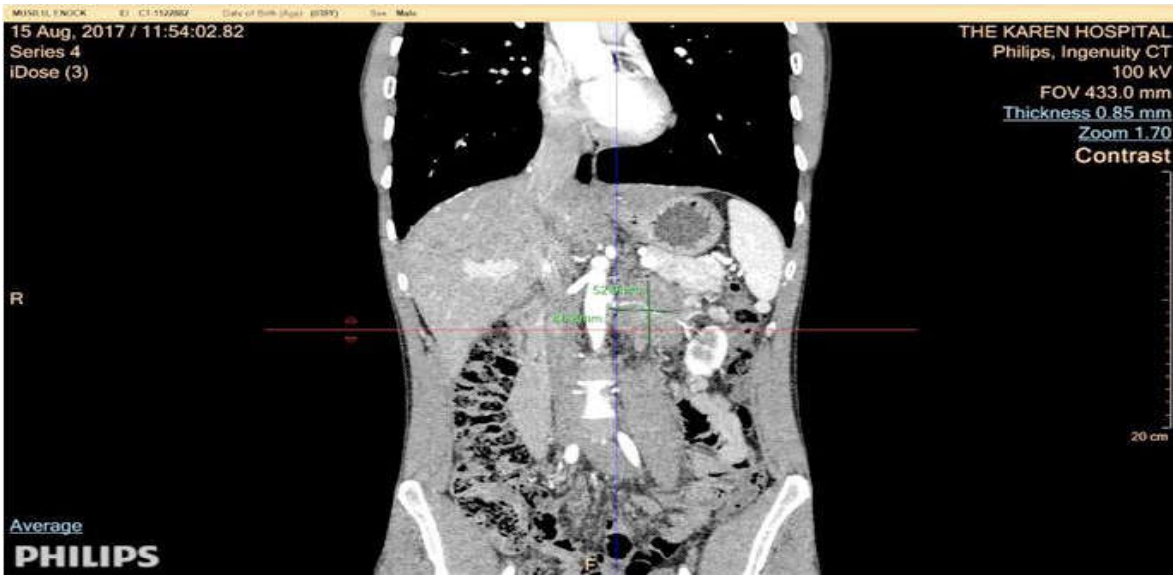

Fig. 1: CT Aortogram Coronal Sectionof patient

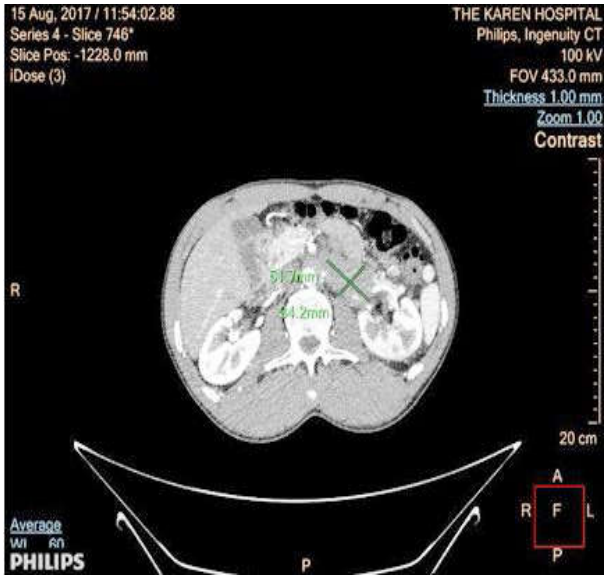

Fig. 2: CT Aortogram Axial view

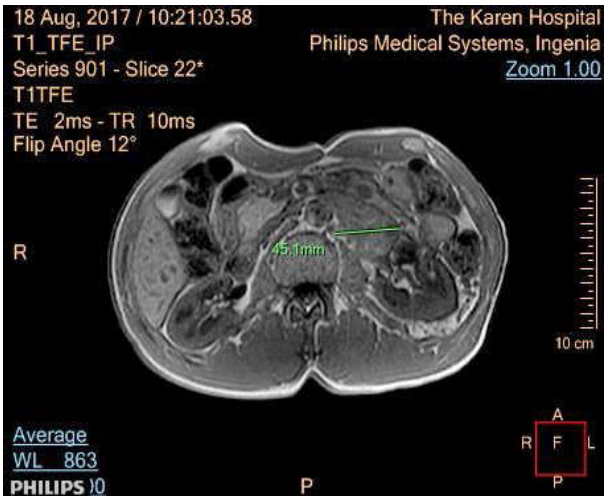

Fig. 3: MRI T1W_TFE Axial View

left kidney. This necessitated for a CT Renal angiogram and aortogram to be done in one sitting, and the following findings were picked.

A hypo-enhancing left renal hilar mass was seen, with encasement and narrowing of the left renal artery. Extension to the aortic origin of the left renal artery. Possible differential diagnoses made were; Retroperitoneal fibrosis, lymphoma or Neuroblastoma.

An MRI abdomen was done which showed an approximately 35mmx49mmx 44mm (APXCCXTR) relatively homogenous ill-defined mass lesion in the retroperitoneum in anterior aspect of the left psoas muscle lateral to the abdominal aorta which is abutted by the lesion along with encasement of the left renal vessels.

BMA Immunohistochemistry

Since lymphoma needed to be excluded out, patient underwent a Bone Marrow Trephinebiopsy. Analysis yielded normocellular marrow, with trilineage haematopoiesis noted. There was no evidence of lymphoma in the bone marrow.

Surgical Management

BMA ruled out Lymphoma, leaving Retroperitoneal Fibrosis and Neuroblastoma as the provisional diagnoses. Decision was made for an excisional biopsy.

Surgery was donevia a chevron incision. Intraoperative findings revealed a retroperitoneal mass encasing the upper pole of the left kidney,

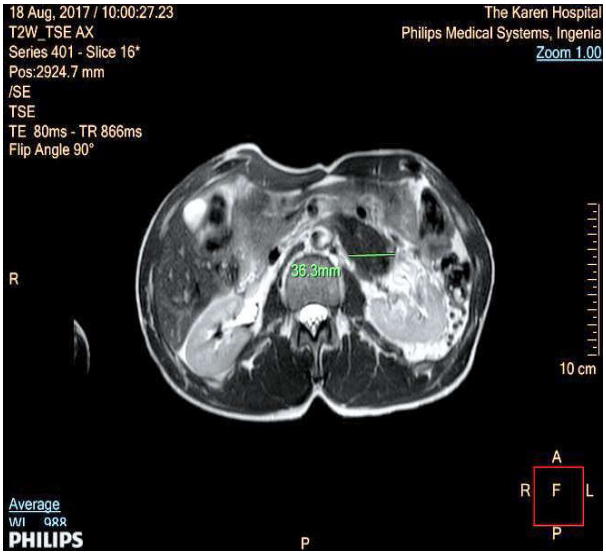

Fig. 4: MRI T2W_TSE axial View

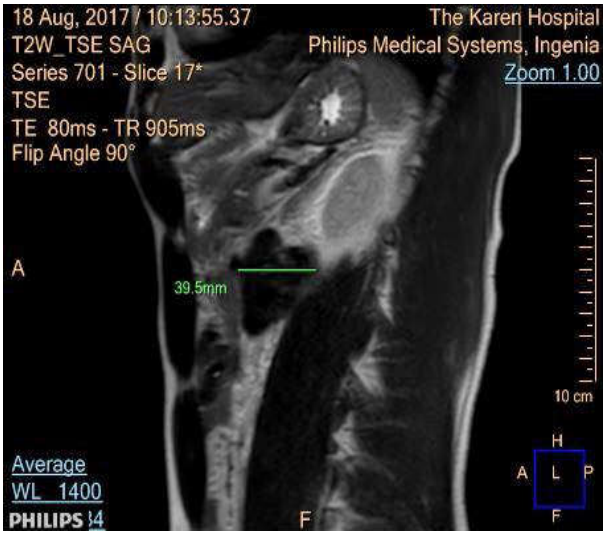

Fig. 5: MRI T2W_TSE Sagittal View

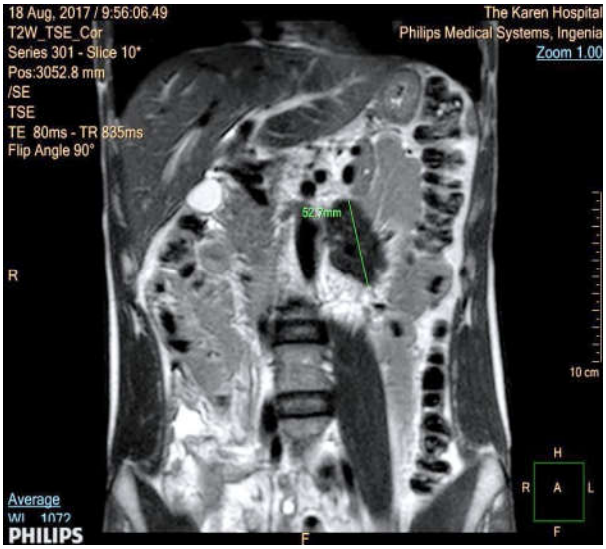

Fig. 5: MRI T2W_TSE Sagittal View

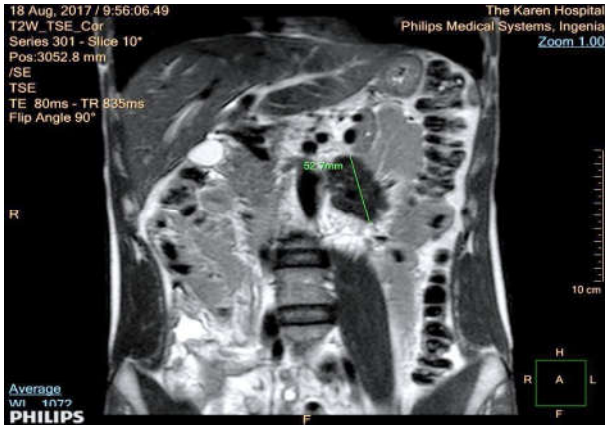

Fig. 6: MRI T2W Coronal View

abdominal aorta medially and the left renal vessels. It measured 45mm by 55mm by 63mm.It was irregular with no clear margins. Decision was made to do a left nephrectomy with abdominal aorta repair. Primary abdominal aortic tissue repair attempts failed: Noted narrowed lumen with poor distal flow. Thus, aprosthetic graft size 22 was used to achieve repair and haemostasis. Patient was then transferred to ICU.

1st day post op he developed right lower limb pain and tenderness. Doppler ultrasound showed right popliteal femoralthrombus. Embolectomy was done.

2nd day Post op, noted intra-abdominal collection with significant drainage from the abdominal drains, warranting for 3rd exploratory surgery to assess graft status.

Intra-operative, large abdominal clots were evacuated with graft status confirmed. Haemostasis achieved. Patient later recovered post op with no new complications after the 3rd surgery. He made a gradual full recovery and was on steroids till pending a histological confirmation. At the time of review after I month he was ambulant, swelling of his legs have subsided, and the BP was under control at 130/82 mm Hg.

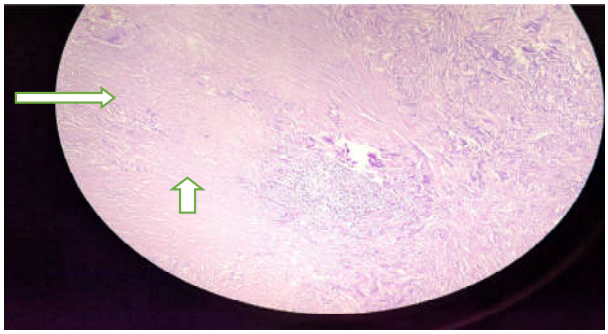

Fig. 7: Microscopic findings of biopsy showing extensive fibrosis

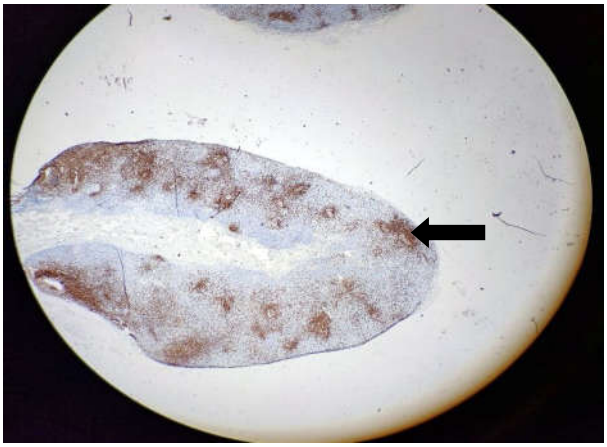

Fig. 8: Immunohistochemistry staining slide

Histopathological Findings: Microscopy

The sections (labelled H0693/17) from the kidney showed no primary pathology.The sections from the Retroperitoneum showed extensive fibrosis and hyalinization. A mixed mature lymphocyte and plasma cell infiltrate was seen.No obliteration of the vessels was evident.

Immunohistochemistry on fibrotic tissue showed a mixture of B & T cells with immunohistochemistry markers CD5 andCD20 respectively.

Final Diagnosis

Retroperitoneum and left kidney:Marked retroperitoneal fibrosis withno evidence of Lymphoma.

Discussion

Retroperitoneal fibrosis is a part of disease spectrum whose common pathogenic process consist of inflammation to advanced atherosclerosis of abdominal aorta. The entities mainly describedare; Idiopathic retroperitoneal fibrosis, Perianeurysmal retroperitoneal fibrosis, isolated periaortitis and inflammatory abdominal aortic aneurysm [1,6,11]. Idiopathic varietyaccounts for two-thirds cases, with the remainder being secondary to drugs such hydralazine, beta blockers, methyl dopa, ergotamine and methylsergide, radiation exposure, asbestos exposure, malignancy; lymphoma, retroperitoneal sarcoma, carcinoid tumours among other metastatic disease from colon, lung, gastric primaries, desmoplastic reaction, iatrogenic retroperitoneal bleeding post-surgery [6-10].

Clinically, retroperitoneal fibrosis has been found to present as an insidious process with initial signs and symptoms being nonspecific, ranging from anorexia, weight loss, malaise, to flank abdominal pain [2,14]. Compression symptoms to ureter, manifests in advanced stages. Laterit presents with features of renal failure; azotaemia, altered level of consciousness, secondary hypertension [1-10]. Signs manifestas peripheral limb oedema from retroperitoneal lymph vessels compression. Venous insufficiency from engorged veins causing varicoceles and hydrocele to deep venous thrombosis [5,8].

Of the above symptoms and signs, our patient manifested hypertension as the key symptom. He illustrated no positive signs of atypical retroperitoneal fibrosis case. This could be explained by the fact that he presented not in advanced stages where the sequel of obstruction from mass effect had not been realised. However, as earlier stated, the symptoms are non-specific and so could provoke thoughts of countless differentials to explain the symptoms. The hypertension was evaluated for possible secondary causes which led the diagnostic team to perform a number of laboratory investigations.

Classical retroperitoneal fibrosis shows acute elevation of inflammatory markers such as C-reactive proteins, ESR in 80 to 100% of patients [19,23,24,25]. Anaemia and azotaemia manifest in kidney injury especially in advanced stages. Rheumatologically tests;(ANA, RF, DsDNA), may be positive [25]. In this case, laboratory investigations were primarily

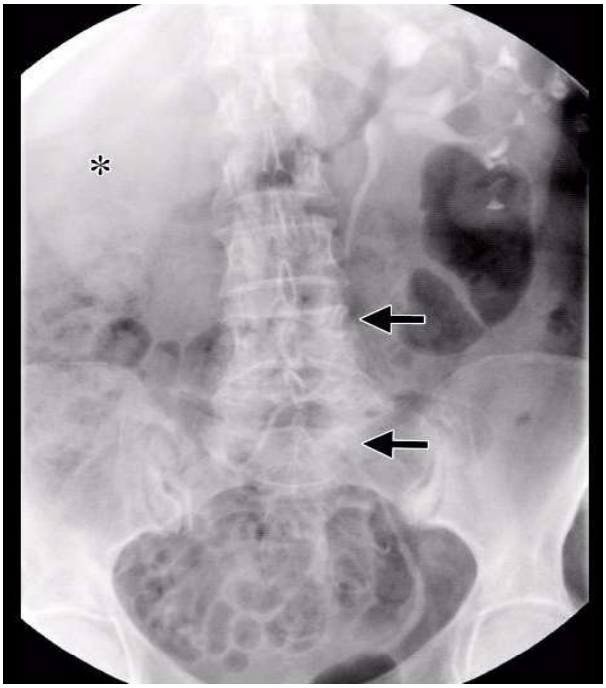

Fig. 9: Urography of Retroperitoneal fibrosis

driven with the need to establish the secondary causes of hypertension such as aldosteronism, Cushing's disease, pheochromocytoma, hyperthyroidism and hyperparathyroidism. Thus not having the differential in mind at the time of carrying out the laboratory evaluation, meant assessment of inflammatory markers and auto-antibodies was overlooked. The laboratory markers yielded no cause of the hypertension and so further radiological investigations were done.

In radiological evaluation of retroperitoneal fibrosis, ultrasound studies yield poor results, with studies showing its poor sensitivity in identification [5,17,18,20]. Normal abdominal X-Ray are also unreliable [18,21]. Intravenous urography and retrograde pyelography is utilized in cases of abnormal kidney function where contrast to patient is a risk [18,22].

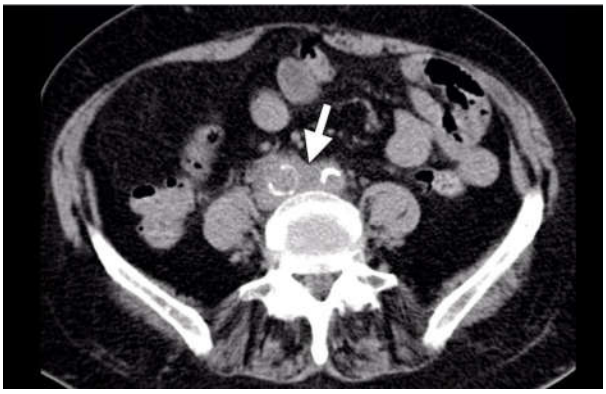

Fig. 10: CT Non Contrast Axial view of retroperitoneal fibrosis

Classical radiographic imaging is as follows;

Its specificity is still low and with advancement in CT guided studies, makes it rarely an option in evaluation [18,22].

CT studies remain the gold standard in obtaining diagnosis in retroperitoneal fibrosis with typical morphology in CT scan showing an isoattenuating to muscle. Below are CT studies images done in other studies for retroperitoneal fibrosis [13,18,21].

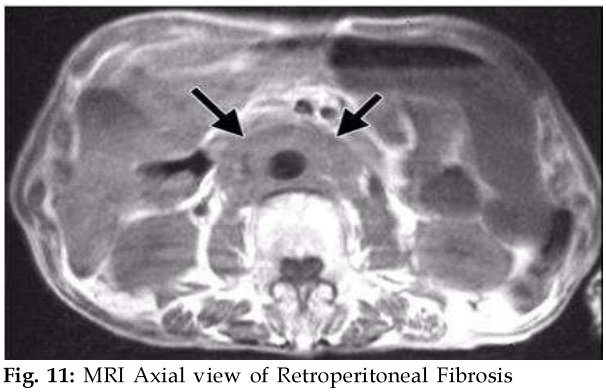

Fig. 11: MRI Axial view of Retroperitoneal Fibrosis

MR imaging has been utilised in diagnosis of retroperitoneal fibrosis with its benefit being eliminating contrast use and without radiation risks of CT studies. High T2 signal intensity is good to identify active inflammation in early disease. For late inactive stages, low T2 signal intensity is important in demonstrating acellular and hypo vascular components of the fibrosis [8,15-19].

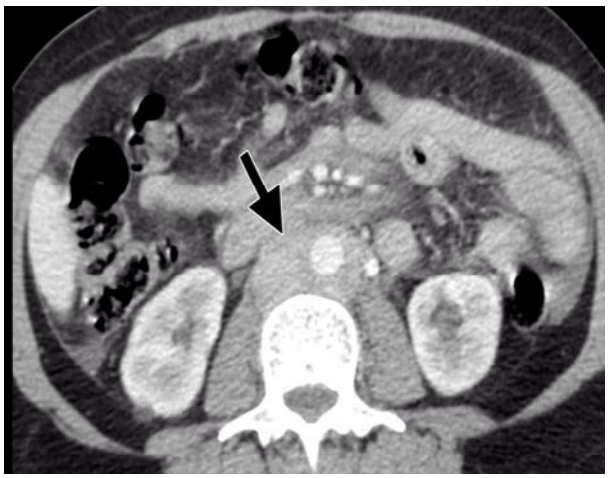

Fig. 12: CT Abdomen Axial view of Non-Hodgkin's lymphoma Case [21]

In our case, we purposed to establish secondary causes of hypertension by starting with a renal Doppler which showed an increased resistance index of left kidney. This necessitated for CT studies that came back diagnostic of a hypo-enhancing left renal hilar mass. This is consistent with studies that show retroperitoneal presentation on CT scans [15-17]. However, since lymphoma was a possible differential, MR studies were done and yielded findings still suggestive of retroperitoneal fibrosis. Though still lymphoma question was not eliminated.

The dilemma of retroperitoneal fibrosis or lymphoma, warranted for a bone marrow aspirate evaluation. It was negative for lymphoma. Though with still worry of missing out early stages of lymphoma despite the negative bone marrow findings, a decision for surgical excisional biopsy was made. Patient underwent surgery but noted post-operative complications as described earlier. Histopathology report showed mass was consistent with retroperitoneal fibrosis. Patient was put on high dose steroid and antihypertensive medication. He progressed well and later discharged.

In the face of radiological diagnostic quagmire, excision biopsy was utilized. Surgery is reserved for

symptomatic relief in advanced stages where mass effect is in play [5,6,9,15,18]. Complications have been documented from postoperative bleeding, to ureteral injuries [9,14,15]. This explains why its use is limited as a last resort in refractory cases.

Summary

Retroperitoneal fibrosis is a rare condition with few incidences which mimic various malignancies. There are only few reports in the world literature on this condition. Radiological investigations still show significant benefit in diagnosis. Excision biopsy has been shown to also offer a diagnosis but with various post-operative complications depicted. More studies are needed fora consensus in guidelines on approach for diagnosis and treatment for this rare entity.

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