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Recurrent Multiple Benign Schwannomas of Left Foot: A Case Report

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Abstract:- Schwannomas are benign neoplasms originating from Schwann cells (1). "It is a soft tissue tumor that rarely presents in the foot (2,3), but in some cases, symptoms typically result from the mass effect and direct involvement of the nerve and surrounding tissue" (1,4). The recommended surgical treatment is tumor Enucleation (1,5–8). We present a case of a 23-year-old man who had recurrent multiple schwannomas of the left foot that were successfully treated with enucleation.

➢ ***Conclusion:***

The most prevalent benign tumors of the peripheral nerve sheath are schwannomas. The recurrence rate after enucleation is around 1.3% to 35.9% in the literature (1,4,25,26). We present a case of recurrent multiple schwannomas in the left foot that were effectively treated with enucleation. Meticulous dissection and complete enucleation are key to decreasing postoperative recurrence and reducing complications.

Keywords:- *Soft Tissue Tumors, Schwannoma, Benign Tumors, Recurrent and Multiple Schwannomas, Enucleation, Microdissection*

I. INTRODUCTION

➢ *Case Report:*

A 23-year-old male patient presented with multiple swellings across the medial aspect of the left foot over a period of one year. The swellings were accompanied by a dull aching pain that subsided with rest. The swelling gradually increased in size over the last 6 months. He was referred to our center by a Primary Healthcare Centre for further management. He was operated the same twice in 2019 and 2020 respectively. No significant history of neurofibromatosis in his family. On physical examination, multiple nodular swellings were present on the medial aspect of the left foot, of about 4cm ×2cm average size. Ultrasonography was suggestive of multiple oval-shaped hypoechoic nodular lesions along posterior tibial and medial plantar nerve. An MRI of the left lower limb revealed multiple oval, atypical soft tissue lesions spreading from the medial side of the left lower leg to the plantar aspect of the foot. On T1W pictures, these lesions were iso hyperintense, but heterogeneously hyperintense on STIR and T2W images. (Figure 1,2,3)

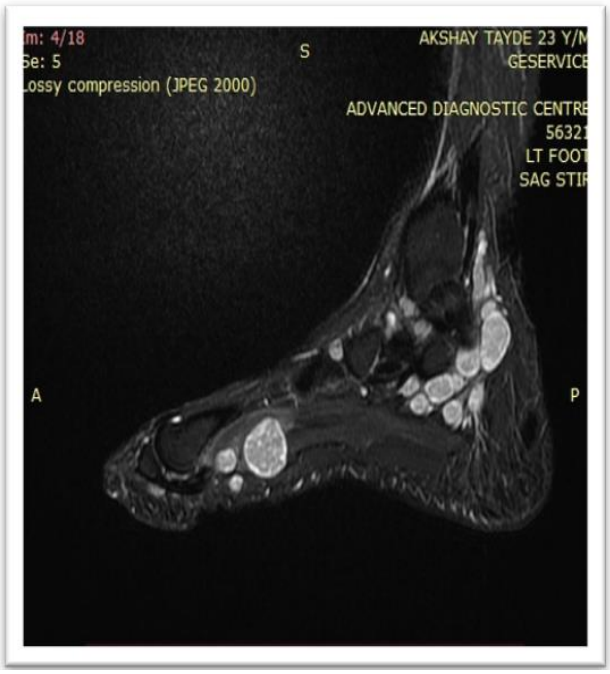

Fig 1:- MRI Saggital View Stir

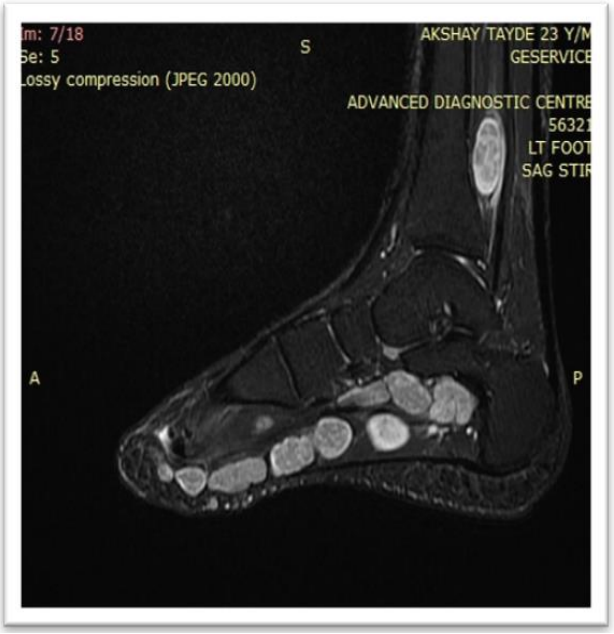

Fig 2:- MRI Saggital View Stir

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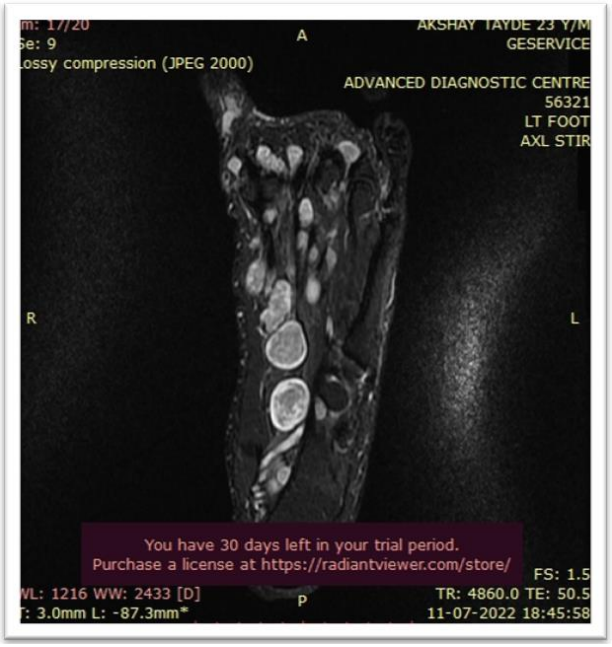

Fig 3:- Axial View Stir of Right Foot

The medial and lateral plantar nerves, as well as their branches, were all affected by these lesions throughout the course of the posterior tibial nerve. The largest of the lesions measured 3.4X1.7cm & 3.3 X 1.7cm on the plantar aspect of the left forefoot. The patient underwent elective surgery an S-shaped incision was made on the medial aspect of the ankle and extended to the sole at the third web space. Multiple schwannomas were identified and enucleated along the course of the posterior tibial nerve, deep peroneal nerve, and medial and lateral plantar nerves, and their branches and the appropriate nerves were preserved. (Fig 4,5,6,7)

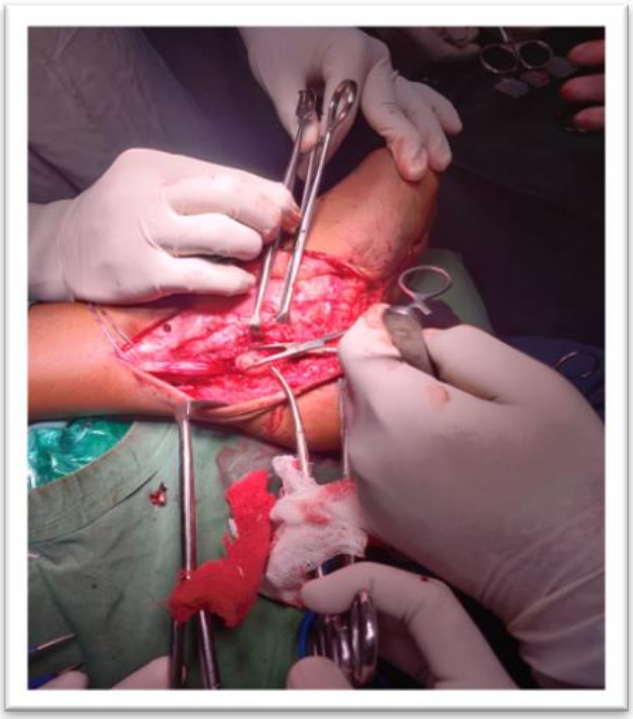

Fig 4:- Along the Deep Peroneal Nerve

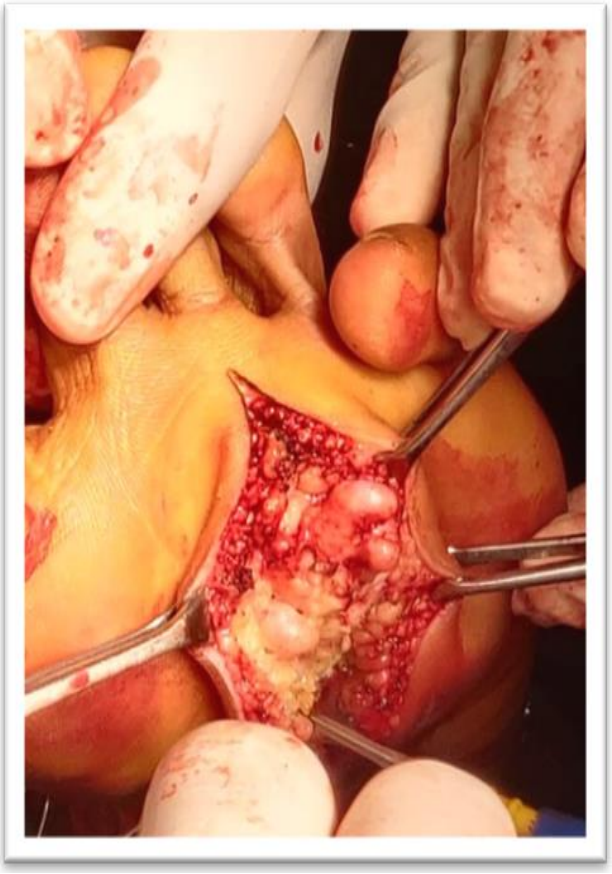

Fig 5:- Sole of the Foot

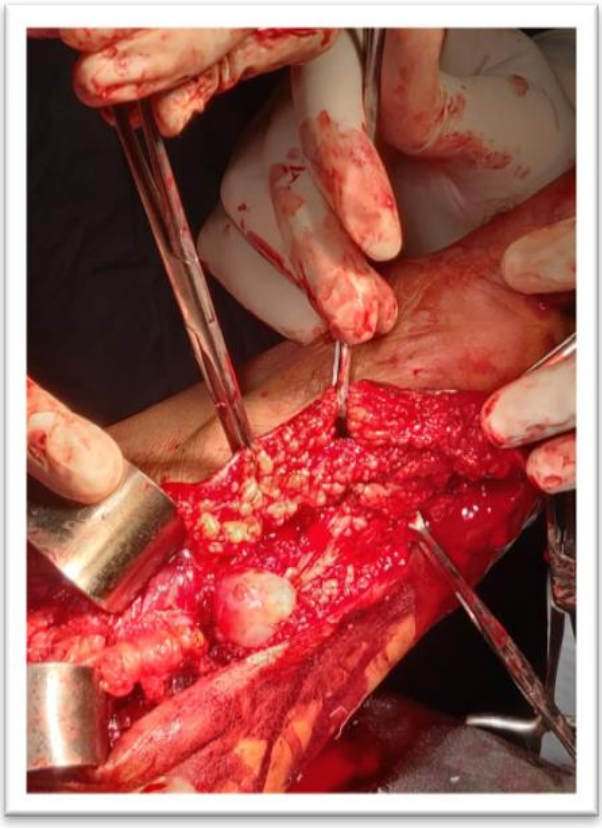

Fig 6:- Along the Medial Plantar Nerve Distally

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Fig 7:- Post-Operative Picture (Multiple Schwannomas)

➢ *Histopathology:*

Gross multiple encapsulated, globular well-circumscribed, solid, homogenous firm, tan glistering tumors ranging from 2-0.3 cm in diameter.

Microscopic examination showed encapsulated, biphasic tumors with alternating hypocellular and hypercellular areas Tumor cells are narrow and elongated, with wavy tapering ends, oval elongated nuclei, and poorly defined cytoplasm. Nuclear palisading and verocay bodies were seen with no evidence of mitosis or malignant changes. Immunohistochemistry showed the lesion was positive for S-100 protein (10,11) which was confirmatory of Schwannomas (figure 8,9)

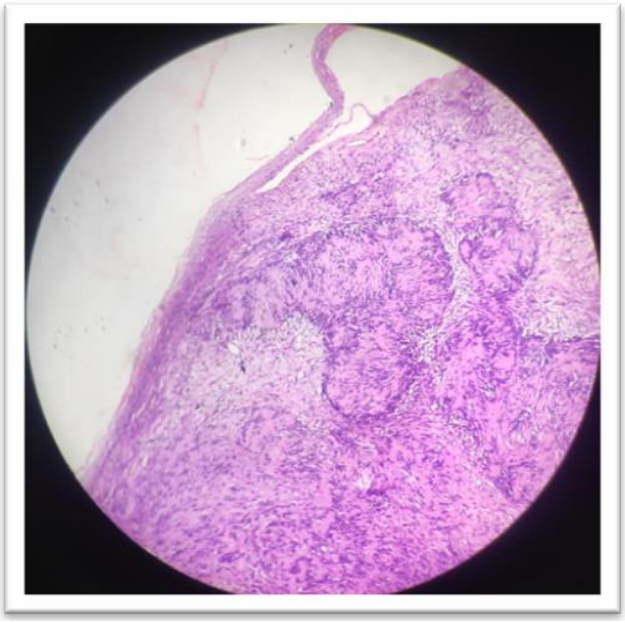

Fig 8:- Encapsulated Tumour

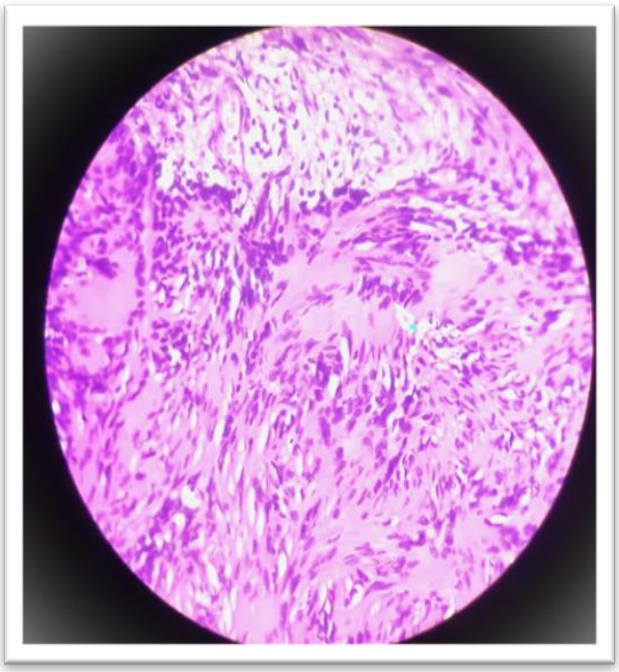

Fig 9:- Verrocay Bodies

➢ *Outcome and Follow-Up:*

The postoperative period was unremarkable, with no symptoms of sensory or motor loss. The patient underwent lower limb physiotherapy and recovered and was discharged on postoperative day 5. The patient is now being followed up 3 monthly for the past 1 year and there have been no clinical signs of local recurrence.

➢ *Discussion:*

Peripheral nerves exit the spinal canal and connect to the upper extremities (arms, hands, and fingers), trunk muscles, lower extremities (legs, feet, and toes), and organs of the body. (12). A nerve's fundamental unit is the axon, which is made up of a cell body, dendrites, and longer axons. All axons are surrounded by Schwann cells, which produce the myelin sheath. (Figure 10)

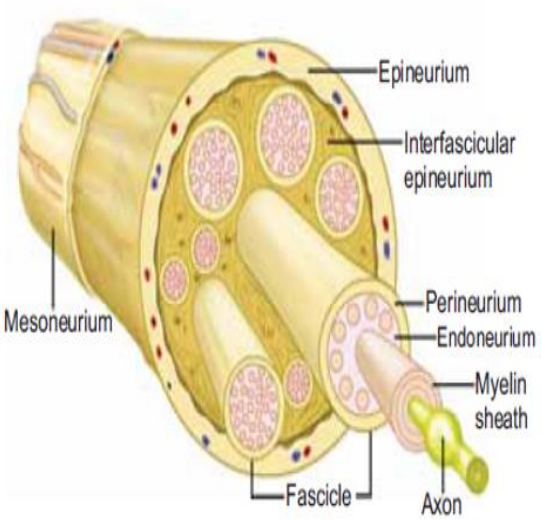

Fig 10

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II. CLASSIFICATION OF PERIPHERAL NERVE SHEATH TUMOURS

➢ *Benign Peripheral Nerve Sheath Tumours*
- Schwannoma
- Neurofibroma
- Perineurioma
- Hybrid Sheath Tumour

➢ *Malignant peripheral nerve sheath tumors*
- Malignant Schwannoma
- Nerve Sheath Fibrosarcoma
- Neurogenic Sarcoma
- Neurofibrosarcoma
- Malignant Neurilemmoma

Schwannomas can occur at any age, although they are more prevalent in people between the ages of 20 and 50, and they affect both men and women equally. They are uncommon in children. It mostly affects the head, neck, and flexor aspects of the limbs, particularly at the elbow, wrist, and knee, as well as the spinal nerve roots, cervical plexus, vagus nerve, and peroneal and ulnar nerves. Lower limbs are seldom affected. Deeply located schwannomas are more common in the posterior mediastinum and retroperitoneum.

Neurofibromas most commonly occur at the age of 20-30 years of age group. The majority are not associated with NF-1 but neurofibromas in NF-1 are more commonly found on the trunk. (1,14,15) table (1)

SCHWANNOMA	NEUROFIBROMA
Consist of only Schwann cells	Both Schwann cells with fibroblast
Focal and Eccentric	Infiltrates along the nerve bundle and Centric
Round	Fusiform
Cysts, Necrosis, and Haemorrhages, are common	Cyst and Necrosis Haemorrhage, are rare
No Malignant degeneration	Malignant degeneration is common

Table 1:- Differnce between schwannoma and neurofibroma

"Hybrid peripheral nerve sheath tumors are most commonly composed of schwannoma-perineurioma combinations, neurofibroma-schwannoma combinations, and neurofibroma-perineurioma combinations" (16). They are commonly associated with NF-1 and have malignant potential; digits are the most common site of presentations.

Six percent of all soft tissue tumors are malignant peripheral nerve sheath tumors. Male to female ratio is 4:1. "The incidence of 0.001% in the general population" (4,17,18). The mean age of incidence of MPNST is 42 years and 70.50% of the MPNSTs are associated with NF-1 and lifetime risk approaches 10%. The proximal lower limb and trunk are the most common sites of occurrence.

➢ *Clinical Presentation of the Peripheral Nerve Sheath Tumors:*

Small schwannomas are typically painless. When the tumor becomes large enough to compress the affected nerve, it might cause paraesthesia, pain, and other symptoms. Pain may migrate along the path of the peripheral nerve. It may be manipulated perpendicularly during a physical examination or during surgery, but not along the nerve's long axis.

In rare instances, Morton's neuroma is a condition that is more commonly seen in females than males in which benign reactive sclerosing process, and is not a tumor originating from Schwann cells (20). It typically originates from the interdigital nerves of the foot, usually the third one. It is probably due to repeated minor trauma. Surgical excision is only performed when necessary.

Neurofibroma presents as a relatively non-tender mass in the skin or subcutaneous tissue. A solitary neurofibroma in the skin usually does not exhibit an associated local change in pigmentation of the skin, although hyper- or hypopigmentation is possible (1,4,14,15).

Pain is the primary symptom of malignant peripheral nerve sheath tumors, which develop from the principal nerves. Sensory and motor deficit such as paraesthesia and weakness are the most common symptoms. (4,17,18)

The diagnostic modalities include radiography, ultrasonography, and MRI. Ultrasound examination of schwannoma often reveals a round or oval, solid, well-delineated, hypoechoic homogenous mass that can be used to differentiate between cystic and solid lesions. "MRI is the best investigation for diagnosis of a schwannoma and MRI signs have been tabulated below" (1,4,18,21) (Table-2)

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	Schwannomas	Neurofibromas	MPNST
T1WI: Homogeneity	Homogenous	More homogenous than schwannomas	25% homogenous 75%Inhomogenous
T1WI: Intensity to muscle	Frequent areas are decreased	Some areas are increased	Increased
T2WI: Homogeneity	More Homogeneous or Inhomogeneous Target appearance may be seen	Less homogenous than schwannomas Target appearance may be seen	Markedly Inhomogeneous Target appearance is uncommon
Necrosis	Frequent	Virtually never	Common
Capsulated	70%	30%	

Table 2:- MRI characteristic features of schwannoma, neurofibroma and MPNST

➢ *Nuclear Medicine:*

When differentiating a benign peripheral nerve sheath tumor from a malignant peripheral nerve sheath tumor using CT or MRI is challenging, nuclear medicine imaging can help. Uptake of 67Ga-citrate occurs in malignant but not benign neural tumors (22).

➢ *Histopathologic Features:*

"Schwannomas are benign, slow-growing neoplasms originating in nerves that are composed exclusively of Schwann cells in the collagenous matrix" (1). Generally, firm, circumscribed, and encapsulated small, spheroidal, larger, ovoid, sausage-shaped. Macroscopic appearance as homogenous tan/grey with irregular yellow areas and cysts. Microscopically there are two types Antoni A, and Antoni B (4,10). "Antoni A compactly arranged spindle-shaped cells with oval, rod-shaped nuclei that arise frequently oriented with a long axis parallel to another creating a pattern of palisades" (10). The distinguishing feature is placed in nuclei rows separated by transparent hyaline bands along the longitudinally sliced bundles known as verocay bodies. Antoni B is mucinous, and microcystic alterations are more common.

❖ *Special Types*

➢ *Cellular Schwannomas:*

Predominantly cellular growth and no verrocay bodies and composed of Antoni A areas. Females are more common than males. It can cause erosion of bone and recurs locally frequently in the retroperitoneum, posterior mediastinum

➢ *Melanotic Schwannomas:*

It is a very rare type. Strong melanocytic differentiation of Schwann cells.50%of the patients develop carney's triad: myxoma, spotty pigmentation, and endocrine activity producing Cushing syndrome.

➢ *Plexiform Schwannomas:*

The most common is seen in males than females. in 5% of all schwannomas, occasionally associated with NF-1 and commonly associated with malignancy

III. TREATMENT

Here we discuss treatment options for benign peripheral nerve sheath tumors Small, asymptomatic tumors may be managed by Close observation for signs of malignancy, including any nerve dysfunction, rapid growth, or rapid increase in pain (1)

The choice of surgery is eventually based on the surgeon's preference

➢ *Pre-Operative Planning:*

MRI is reviewed to confirm the anatomy of the tumor and the characteristics and planning of the surgical approach.

Nerve reconstruction options are discussed with the patient.

➢ *Surgical Approach:*

- *Enucleation:*

Most isolated BPNSTs are schwannomas and arise eccentrically from the nerve sheath. Tumours are encapsulated and can be safely enucleated without removing the nerve fascicle (1,6–9)

Inspect the nerve circumferentially for the window of splayed fascicles that affords the best resection plane. The resected specimen should contain no nerve fascicles.

- *Microdissection:*

Digital nerve schwannomas may sometimes require microdissection (1,5,23) to preserve axons. The tumors are isolated under loupe magnification by the surgeon to identify and preserve the normal fascicles.

The Nerve Stimulator will allow for checking of motor function intraoperatively and, allow for a safer dissection of these small challenging tumors.

➢ *Post-Operative Care:*

Postoperatively a minimum, short arcs of joint motion are initiated early. If necessary, the end range of motion may be avoided for up to 1 month to protect nerve repairs or reconstructions (24).

➢ *Outcome:*

Transient paraesthesia is common after the enucleation of schwannoma.
Microdissection preserves the nerve function, but it has an increased risk of recurrence

➢ *Complications:*

Pain is the most common complication. Loss of nerve function after tumor resection due to extensive dissection can occur. Loss of motor function due to prolonged immobilization is also a rare complication. (1,25)

IV. CONCLUSION

The most prevalent benign tumors of the peripheral nerve sheath are schwannomas. The recurrence rate after enucleation is around 1.3% to 35.9% in the literature (1,4,25,26). We present a case of recurrent multiple schwannomas in the left foot that were effectively treated with enucleation. Meticulous dissection and complete enucleation are key to decreasing postoperative recurrence and reducing complications.

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