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# Case Report of Diffuse Neurofibroma in the Right Supraclavicular

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**Abstract** **Case Report**

The diffuse neurofibroma is a rare but particular form of neurofibroma, mainly occurring in children and young adults. About 10% of patients with diffuse neurofibroma have neurofibromatosis type 1. Due to its rare association with neurofibromatosis, it is a rare and difficult diagnosis. Its imaging appearance is very similar to that of a vascular malformation, and it is often misdiagnosed until pathological study proves otherwise. We report the case of a 48-year-old patient who complained a mass in the right supraclavicular region, with imaging suggesting a hemangiopericytoma. However, it was later found to be a diffuse neurofibroma.

**Keywords:** Diffuse neurofibroma, Neurofibromatosis type 1, Soft tissue tumor, Magnetic resonance imaging, S100 protein, Surgical excision.



## Background

Soft tissue neurofibromas represent approximately 5% of all benign soft tissue tumors in large surgical series, they are usually divided into three different types: localized, plexiform, and diffuse, with most commonly affecting patients aged 20 to 30 years old. Among these, the diffuse variety is the least common and it's most often confused with a vascular malformation due to its close imaging appearance. Diffuse neurofibromas affect the skin, and subcutaneous tissues of the head and neck region, trunk, and extremities.

## Case Presentation

We report the case of a 48-year-old patient who consulted for a 6.5-centimeter, painless and pedunculated mass, located in the right supraclavicular area (Figure 1). It had been evolving since the patient was 26 years old and had gradually increased in size. The overlying skin appeared normal, without any inflammatory signs. Clinical examination hasn't revealed any cervical lymph nodes or light-brown spots on the trunk or extremities. There have been no similar familial cases, and the rest of the clinical examination was normal. The patient had a CT scan, which revealed an exophytic mass, in the right cervical area, with a hypodense (Figures 2,3) appearance and enhancing after contrast injection. The mass contained many vascular structures. Magnetic resonance imaging (MRI) was performed, and it revealed a well-defined mass with regular contours in the subcutaneous tissue of the right supraclavicular area. It appeared hyperintense on T2-weighted images, hypointense on T1-weighted images (Figures 4,5), non-restrictive on diffusion sequences, and showed intense and heterogeneous enhancement after contrast. The mass respected the deep muscular plane and measured 57 mm in height, 75.5 mm in transverse diameter, and 64 mm in anteroposterior diameter, initially suggesting a diagnosis of hemangiopericytoma, or a possible dermatofibroma.

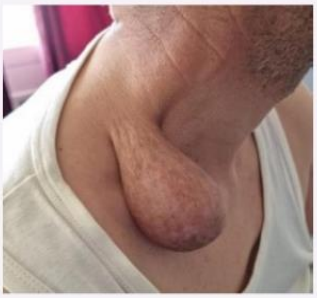

Figure 1 Clinical Image of the supraclavicular mass.

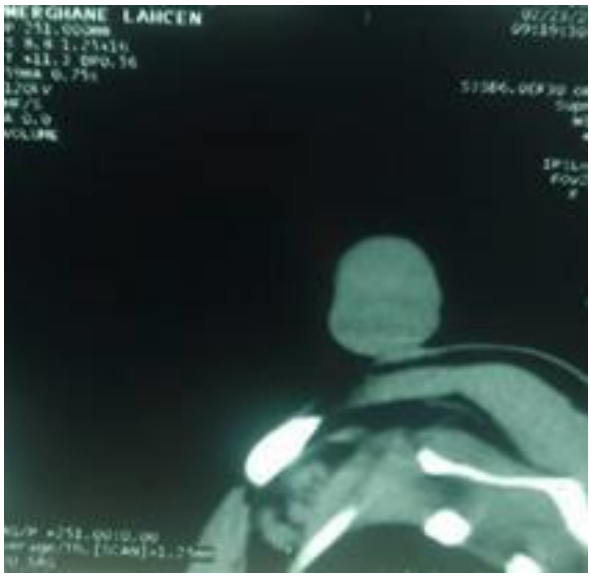


**Figure 2**: **CT Scan images show a hypodense mass compared to the muscle.**

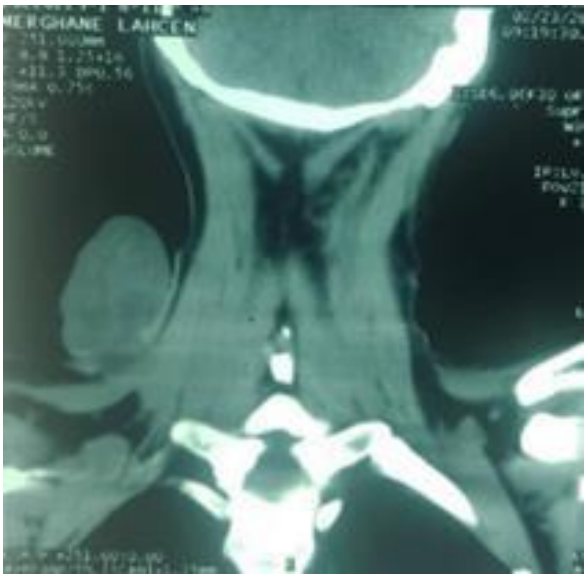


**Figure 3: CT Scan images show a hypodense mass compared to the muscle.**

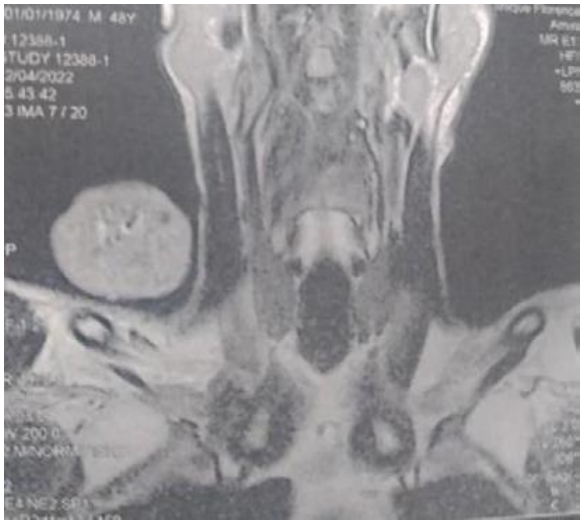

**Figure 4: MRI images show a mass in hypersignal T2 and hyposignal in T1.**

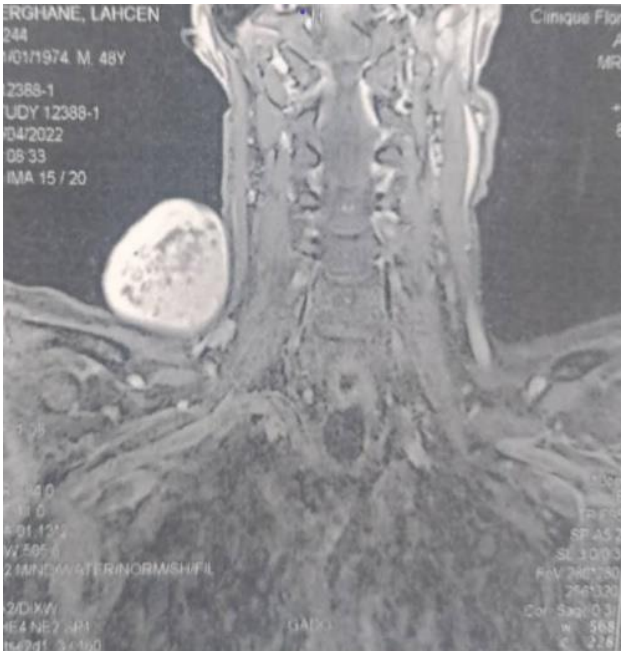

**Figure 5: MRI images show a mass in hypersignal T2 and hyposignal in T1**

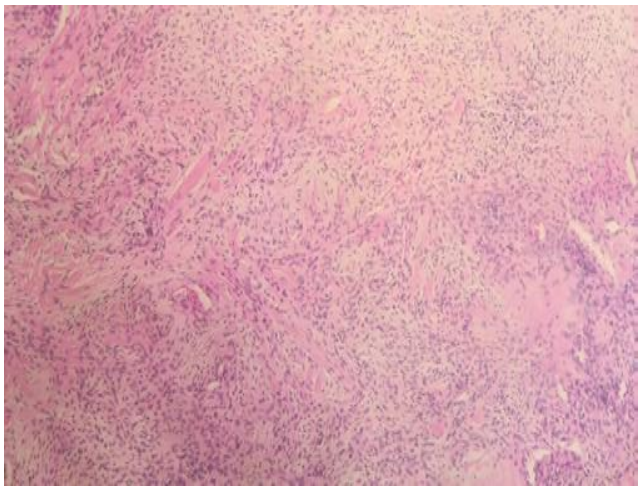

**Figure 6: Neurofibroma: tumor proliferation made up of non-atypical spindle cells arranged on a fibrous background (H&E x200).**

**Figure 7: Neurofibroma: immunohistochemical study shows expression of S100 protein by the tumor cells.**

However, the duration of disease progression and the symptomatology did not suggest malignant pathology.

A mass excision was performed with a 1 cm margin of safety at the base of implantation. Histopathological analysis of the specimen showed spindle or oval-shaped tumor cells with generally regular nuclei, slightly increased in size, with fine chromatin and eosinophilic cytoplasm. Mitoses were rare. The proliferation contained numerous dilated vascular structures with cystic cavities bordered by flattened cells. Immunohistochemistry showed positivity for S-100, consistent with a diagnosis of diffuse neurofibroma (Figure 6,7).

## DISCUSSION

Schwannoma and neurofibroma are common benign tumors of the nerve sheath. Neurofibroma represents about 5% of all benign tissue tumors in large surgical series and is generally divided into three different types: localized, plexiform, and diffuse [1,2,3] Malignant transformation of neurofibromas is rare (Figure 6). The risk is increased if associated with neurofibromatosis, especially with the plexiform type [4,5]. Malignant peripheral nerve sheath tumors (MPNST) can occur in 2% to 13% of patients with NF type I, compared to 0.001% of the general population [6,7]. The signs of malignant transformation are mainly the painful character and rapid growth of the mass. Diffuse neurofibroma is a rare subtype of neurofibroma that mainly affects children and young adults [8,9]. It usually affects the skin and subcutaneous tissue of the head and neck, trunk, and extremities. It's a benign tumor with rare malignant transformations [10-12]. At least 10% of these tumors are associated with NF-1 [13,14] although Megahed reported 61% of his 13 patients with diffuse neurofibroma to have NF-1 [15]. Due to lack of established imaging criteria, the radiological description of diffuse neurofibroma is unclear in the literature [16,17]. Diffuse neurofibroma is most often confused with a vascular malformation due to its close imaging appearance [1 ,9,18]. On CT scan, pre-contrast images appear hypodense. Marked hyperdensity of the tumor on contrast-enhanced scans is indicative of its rich vascularity [19]. On magnetic resonance imaging (MRI), most of these lesions are described as isointense or slightly hyperintense to muscle on T1-weighted images, and slightly or markedly hyperintense to muscle on T2-weighted images. Such findings have been consistently described in various case reports [8,20]. Histologically, it is marked by diffuse replacement of the dermis by bundles of cells with round or fusiform nuclei and eosinophilic cytoplasm, within a matrix of fibrillary collagen [21]. Despite the remarked infiltration, it does not destroy tissues but envelops and entraps the normal structures, such as skin adnexal structures, adipose tissue, or muscle fibers. Meissner corpuscles are characteristic of diffuse neurofibroma, they are observed in almost all cases. However, Meissner corpuscles are rarely missing, as reported in a few studies [21]. Diffuse neurofibroma is different from dermatofibrosarcoma protuberans, which shows no S-100 positivity and has no Meissner corpuscles [22,23]. The treatment of large neurofibromas consists of partial or complete surgical excision [24-25]. In some cases, angiography and intra-arterial embolization can be performed preoperatively, because of troublesome bleeding that may occur during excision [24,26]. Even after complete excision, diffuse neurofibromas may recur because of their infiltration [2,9]. Due to the possible recurrence and potential development of neurofibromatosis, annual follow-up is recommended.

## CONCLUSION

Diffuse neurofibroma is a rare subtype of neurofibroma and is often not associated with neurofibromatosis. Moreover, its imaging appearance is unclear and may lead to misdiagnosis as a vascular malformation. Therefore, knowing this pathological entity and its radiological characteristics leads to a better diagnosis and appropriate therapeutic management.

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