

LARGE SOLITARY ENCAPSULATED NEUROFIBROMA OF UPPER ARM – A CASE REPORT

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Abstract: Introduction: Neurofibromas are benign tumors of neuronal origin, occurring most commonly in young adults, with no gender predilection. The connection of neurofibroma with disorders on a general level as von Recklinghausen's disease makes its diagnosis critical. **Case report:** A 32-old female patient was administered to the Clinic of Plastic surgery, Clinical Center of Montenegro in Podgorica, with a 10-year history of painless, subcutaneous tumor of the right upper arm that grew in size over the last ten years. The patient reported progressive pain and tingling in her right forearm and right hand for the last 12 months. An MRI showed a non-homogenous tumor of the middle third portion of the triceps muscle, in close contact with the humerus but without infiltrating it. The tumor was removed, with a definitive histopathological result of a solitary benign neurofibroma. **Conclusions:** This example of successful treatment of solitary neurofibroma may serve to increase the awareness of surgeons and radiologists in small countries regarding benign peripheral nerve sheath tumors. The patient is under observation for two years with no signs of relapse and no other features indicative of neurofibromatosis type 1.

Keywords: peripheral nerve sheath tumors, neurofibroma, magnetic resonance imaging, surgery, upper arm.

INTRODUCTION

Neurofibromas are benign peripheral nerve sheath tumors that can develop as solitary tumors or as a part of neurofibromatosis type 1 (NF-1). We report an uncommon case of a large solitary neurofibroma with no neurofibromatosis type 1 presence. Radical surgery remains the choice of treatment in such cases, providing low recurrence and the best long-term results.

CASE REPORT

A 32-year-old woman presented to the Clinic of Plastic surgery Clinical Center of Montenegro in Podgorica, with a palpable subcutaneous tumor of the right upper arm. She stated that the mass grew slowly over the last ten years, without any symptoms until last year when periodical tingling of the forearm and right hand (thumb) pain occurred. The symptoms worsened in the recent couple of months. Her previous medical history and laboratory results were unremarkable, and the patient denied a family history of NF-1. The clinical examination revealed in the middle portion of the right upper arm an irregularly-oval-shaped subcutaneous tumor, approximately 6 cm in its diameter. It was palpably painless, there wasn't a vascular deficit on that arm, and Tinel's sign on the median nerve was positive. An magnetic resonance imaging (MRI) of the right arm showed an irregularly oval-shaped, non-homogenous, necrotic tumor measuring 70 x 36 mm, with extension to subcutis in one direction, and in other close contacts with the cortex of humerus, without infiltration of the bone in the middle third of the triceps muscle (Figures 1 and 2). No detectable vascular or bone injuries and associations were seen on the MRI. The patient was examined for clinical signs of NF-1, and from a minimum of two of seven suggested criteria for confirming the diagnosis of NF-1, in our patient all criteria were negative (explained in further text). Then, under general anesthesia, the extirpation of the tumor was performed. Surgery revealed an irregularly dome-shaped tumor measuring 60x53mm, with a smooth surface and skin-like color, with medium firm consistency (Figure 3), located in the middle portion of the triceps muscle, with no visible infiltration of adjacent bone and muscle tissue. The tumor was removed and subjected to definitive histopathological examina-

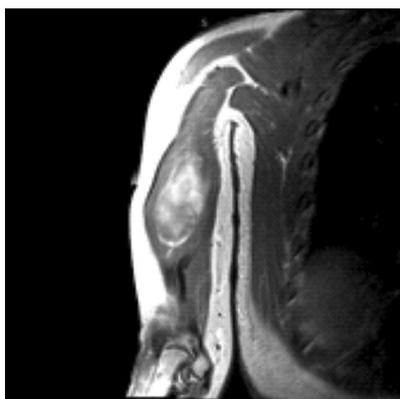


Figure 1. Sagittal plane MRI: showing tumor in the right upper arm

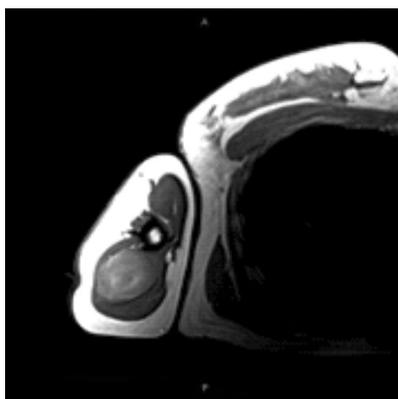


Figure 2. Transversal plane MRI: showing tumor in the right upper arm

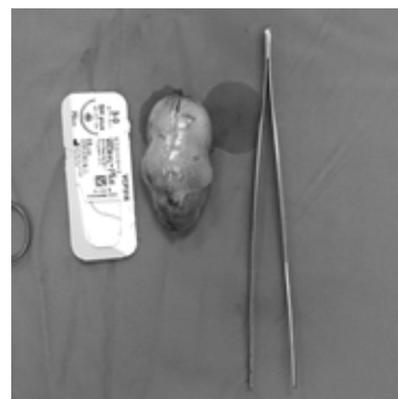


Figure 3. Extirpated tumor

tion. The postoperative period was uneventful, without relapse of the tumor. Histopathology reported regular histological and cytological built - a tumor made of spindle-shaped fibroblasts and Schwann cells, normochromic wavy nuclei, with no mitosis included in the mostly hyalinized and partially myxoid stroma, encapsulated by a thin capsule. The further immunohistochemical analysis stated s-100 positive, neuron-specific enolase, vimentin, and negative immunoreactivity to pan-cytokeratin and desmin. All this considered, the tumor was defined as a benign neurofibroma, without evidence of neurofibromatosis.

DISCUSSION

Clinically, neurofibromas arise in two possible patterns: either as sporadic tumors or in association with neurofibromatosis, Von Recklinghausen's disease.

Neurofibromatosis, Von Recklinghausen's disease is a direct consequence of a defect on chromosome 17, encoding for a tumor suppressor gene NF1, which is transmitted in an autosomal dominant pattern. It affects the skin, nervous, musculoskeletal system, and eyes. Two of the seven criteria are required to confirm the diagnosis of NF-1 (1): six or more café au lait macules, freckling in the axillary or inguinal lentiginos, two or more neurofibromas of any type or a plexiform neurofibroma, two or more Lisch nodules (iris hamartomas), osseous lesions, an optic glioma and an affected first-degree relative (sibling or parent) (2, 3, 4).

Histologically three subtypes of neurofibroma are: solitary, diffuse, and plexiform. Solitary, sporadic neurofibromas are most common, with no racial and gender predilection, most often occurring in adults in the third decade. They present as usually slowly growing, skin-colored, rubbery tumors (5). Diffuse neurofibromas are most common in children, usual-

ly located within the subcutaneous tissue of the head and neck. Plexiform neurofibromas are characterized by diffuse involvement of the nerve segment and its branches, often accompanied by massive soft tissue overgrowth and consequently functional impairment. They are pathognomonic of NF-1, and unlike solitary and diffuse types, the plexiform type is associated with an increased risk of malignant transformation.

Solitary neurofibromas are usually clinically silent at the beginning, but as they inchmeal increase in diameter, compression effects occur on adjacent structures and organs, which exactly had happened in our patient, with Tinel sign positive. Giant variants of solitary neurofibroma exceeding 2 cm in diameter are rare, and those which are not associated with type I neurofibromatosis (NF-1) are even rarer. Such cases were reported in head & neck (6, 7), preperitoneal (8), retroperitoneal (9, 10), oral cavity (11), thoracic cavity and extremities.

The exact etiopathogenesis of solitary sporadic neurofibroma is still unknown, with the most acceptable theory considering it as a hyperplastic hamartoma (12, 13, 14).

Preoperative imaging (ultrasound, CT, MRI) is often inadequate to provide a definitive diagnosis, although data received from preoperative imaging facilitates the surgeon's job (ex. to determine the general morphology of the tumor, its location, and correspondence with adjacent structures) and therefore helps to prevent intraoperative complications.

When possible, if an incisional biopsy is performed with consequent pathohistological confirmation of neurofibroma, opinions concerning further treatment options are divided between clinicians. Few authors argue that only when the tumor is a cause of neurological deficiencies and/or its malignant potential is suspected radical surgery indicated (15). Authors of this text believe that if there are no vital con-

traindications, surgical excision in toto should always be performed. Besides functional impairment, solitary neurofibroma can also destructively engage adjacent structures, leading to irreversible loss of function in damaged structures, leading to worse outcomes, such as amputation of the limb (16).

A definitive diagnosis can only be provided by a pathologist after conventional histological analysis and immunohistochemistry. Histopathological examination confirms the proliferation of Schwann cells, perineural cells, and fibroblasts are seen among the stroma, sometimes myxomatous and micro -vacuolation. Differentiating neurofibroma from schwannoma is focal S-100 positivity which is a characteristic of neurofibroma. Histopathological findings show proliferation of Schwann cells, perineural cells, and fibroblasts amid a myxomatous stroma. The differential diagnosis for painful subcutaneous tumors should include myxoma, neurofibrosarcoma, angioliipomas, rhabdomyoma, and especially schwannomas, with preoperative imaging often being insufficient in differentiating them (17, 18).

Sažetak

VELIKI SOLITARNI NEUROFIBROM NADLAKTICE-PRIKAZ SLUČAJA

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Uvod: Neurofibromi su benigni tumori nervnog tkiva, javljaju se najčešće kod mlađih odraslih osoba, podjednako kod oba pola. Potencijalna povezanost neurofibroma sa sistemskim poremećajem poput neurofibromatoze tip 1 (von Recklinhausen-ova bolest) čini dijagnozu neurofibroma izuzetno značajnom. **Prikaz slučaja:** Prikazujemo 32-godišnju pacijentkinju sa prisutnom potkožnom tumefakcijom u predelu desne nadlaktice, koja je prisutna unazad oko 10 godina, uz postepen rast. Pacijentkinja se žali na osećaj bola i trnjenja u desnoj podlaktici i šaci unazad oko 12 meseci, sa progresivnim pogoršanjem tegoba. MR dijagnostika potvrđuje prisustvo nehomogenog tumo-

CONCLUSION

Regarding soft tissue tumors, especially in young adults, it is recommended to include neurofibromatosis as part of differential diagnosis. A follow-up is fundamental for younger adult patients who were diagnosed with a solitary neurofibroma so that diagnosis of NF-1 can be excluded with certainty. In our patient, the absence of distant metastases, the absence of required signs of NF-1, as well as no relapse after surgery, and uneventful follow-up - confirmed the nature of the tumor.

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ra u predelu srednje trećine tricepsa desne nadlaktice, koji je u bliskom kontaktu sa humerusom, ali ga ne angažuje. Tumor je ekstirpiran u celini, sa histopatološkom verifikacijom solitarnog benignog neurofibroma. **Zaključak:** Ovaj prikaz slučaja uspešnog lečenja solitarnog neurofibroma služi kao podsticaj za podizanjem svesti za neurofibrome i tumore omotača perifernih nerava među hirurzima i radiolozima u manjim centrima. Pacijentkinja je redovno kontrolisana tokom dve godine postoperativno, bez recidiva i znakova razvoja NF-1.

Cljučne reči: tumori omotača perifernih nerava, neurofibrom, magnetna rezonanca, hirurgija, nadlaktica.

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