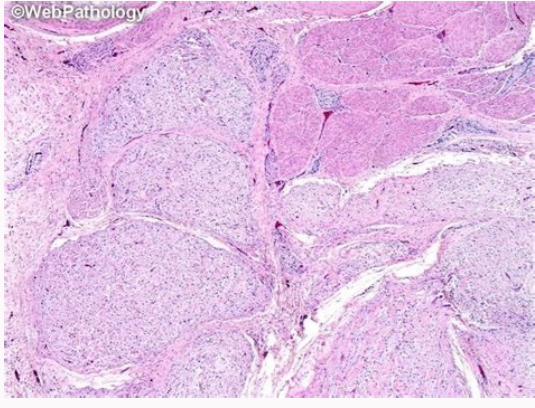
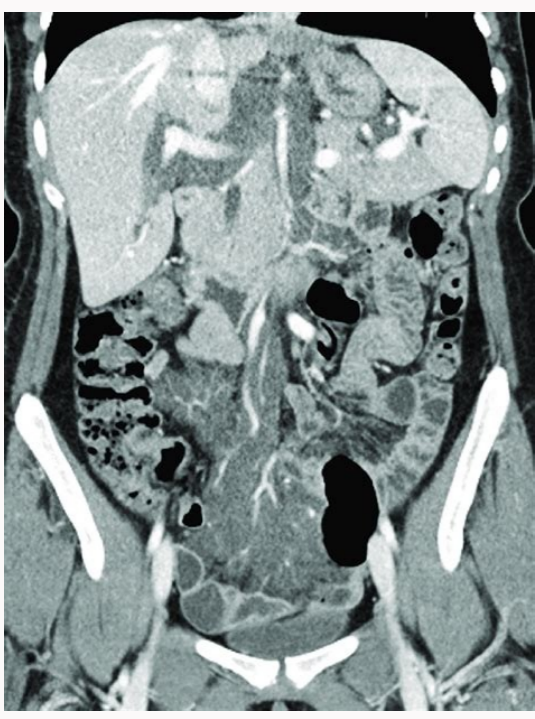




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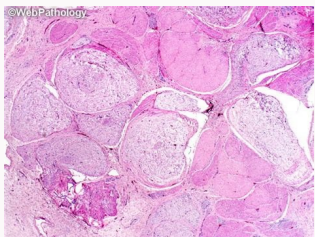



# PLEXIFORM NEUROFIBROMA

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These tumors are known to cause symptoms ranging from minor discomfort to extreme pain. Two types of plexiform neurofibromas have been recognized: (a) a diffuse type/ elephantiasis neurofibromatosa and (b) a nodular type (4, 5). These plexiform neurofibromas can arise anywhere along a nerve and have poorly defined margins. 2nd edition. The cranial nerves most commonly involved in plexiform neurofibromas are the fifth, ninth, and tenth. Areas of freckling and regions of hypertrichosis occasionally overlay plexiform neurofibromas. J Med Genet. It is a virtually pathognomonic and often disabling feature of neurofibromatosis type 1. A clinical study of type 1 neurofibromatosis in northwest England. There was scoliosis of the cervical spine. They are nodular and may be brown, pink, or skin-colored and soft to firm to touch. They originate from nerve-sheath cells or from subcutaneous or visceral peripheral nerves and can involve multiple fascicles. Bethesda, Md., USA, July 13-15, 1987. The term "plexus" refers to a combination of interlaced parts or a network. This architecture could account for the centrally T2 dark (nerve fibers) and peripherally T2 bright (myxoid) appearance of these target-like lesions. [PubMed] [PMC free article] [PubMed] [Google Scholar] Articles from Radiology Case Reports are provided here courtesy of Elsevier 1999; Mar 26;89(1):31-37. The consistency of the lesion has been compared to that of a "bag of worms" because of the presence of soft areas interspersed with firm nodular areas. Sexual precocity is seen in 3-5% of affected children. Facial plexiform neurofibroma may produce various degrees of cosmetic and functional deformities in the head and neck region. This freckling is not seen in patients without neurofibromatosis. Extension of the lesion was seen posterior to the back of neck and up to the right paravertebral region. MRI showed overgrowth of the skin and subcutaneous tissue that was hyperintense on T2-weighted images (Figure 5, Figure 6). Incisional biopsy was performed under general anesthesia. These lesions sometimes demonstrate a vascular nature, and they may cause dangerous bleeding and complicate surgical procedures. [PubMed] [PubMed] [Google Scholar] 4. Cutaneous and subcutaneous varieties are not specific for neurofibromatosis, whereas the plexiform variety is specific for the disease. Roughly 20-30% of patients with neurofibromatosis have axillary freckling, known as Crowe's sign. There were dysplastic changes and deformity involving the right side of maxilla, mandible, and upper cervical vertebrae. There was no history of any regression in size of the swelling or any discharge from the swollen area. Plexiform neurofibroma is a rare, poorly defined benign tumor of the peripheral nerve sheath. The patient had never noticed similar swelling elsewhere in the body. Johns Hopkins University Press; Baltimore: 1992. It has one of the highest spontaneous mutation rates among genetic diseases. It is an autosomal dominant disorder caused by a spectrum of mutations in the NF-1 gene. Family history did not reveal any similar complaints in immediate or distant relatives. The patient underwent several imaging procedures. Plexiform neurofibromas are uncommon and occur almost exclusively in about 5-15% patients with neurofibromatosis-1. These masses can be quite disfiguring, and hemifacial hypertrophy can occur (6). It spreads out just under the skin, or deeper in the body, and occurs exclusively in patients with neurofibromatosis type 1. Pathology of tumours of the peripheral nerve sheath in type 1 neurofibromatosis. This consistency was well appreciable in the lesion seen in our patient. Superficial plexiform neurofibromas do not typically possess the target-like appearance seen in their deep counterparts and can be easily mistaken for other entities, such as venous malformations. Only 50% of these patients have a positive family history of the disease; the other half represent spontaneous mutations (1). Other possible abnormalities that may be seen include central nervous system tumors, macrocephaly, mental deficiency, seizures, and short stature (2). Am J Med Genet. 1999 Mar;36(3):197-203. Nodules (Lisch nodules) and café au lait macules of the iris bilaterally have been described as characteristic of neurofibromatosis in the ophthalmological literature (3). Plexiform neurofibromas are benign tumors that spread out either just under the skin or deeper in the body. Woodruff JM. [PubMed] [PubMed] [Google Scholar] 5. [PubMed] [Google Scholar] 3. The patient first noticed the swelling ten years ago; it began insidiously and then grew slowly to attain its present size (Figs. At least eight forms of neurofibromatosis have been recognized, the most common form being neurofibromatosis type I (NF-1), also known as von Recklinghausen's disease, which accounts for about 90% of the cases. Friedman JM, Birch PH. Our patient had numerous café au lait spots and one plexiform neurofibroma. Neurofibromas are the most common benign tumors of NF-1. Both axillary freckling and inguinal freckling may develop during puberty. Neurofibromatosis. Computed tomography (CT) showed overgrowth of the skin and subcutaneous tissues, with invasion and encasement of the muscles and soft tissues of the right side of the face (Figure 2, Figure 3, Figure 4). These can develop at any point along a nerve and often form by late adolescence. [PubMed] [PubMed] [Google Scholar] 6. 1A, 1B) After an asymptomatic phase of nine years, the patient began to experience mild, intermittent, dull, aching pain in the swollen area, which continued over the past year. McCaughran JM, Harris DI, Donnai D. National Institutes of Health Consensus Development Conference Statement: Neurofibromatosis. Riccardi VM. We present a case of plexiform neurofibroma in an 18-year-old female. Abbreviations: CT, computed tomography; MRI, magnetic resonance imaging. An 18-year-old female visited our diagnostic center with a chief complaint of a swollen area on the right side of the face. 1997; May 16;70(2):138-143. The absence of a target-like appearance does not rule out a lesion being a neurofibroma, particularly if the lesion has a superficial location. Published: September 24, 2012. Type 1 neurofibromatosis: a descriptive analysis of the disorder in 1,728 patients. 1988;1(3):172-178. Deep plexiform neurofibromas consist of a nerve or nerve fascicle distended by tumor cells, embedded in a rich myxoid matrix. The size of these tumors may increase during puberty and pregnancy. Deep plexiform neurofibromas typically present with a target-like appearance on T2-weighted MR images, with central low-signal intensity and peripheral high-signal intensity. Longitudinal bundles of residual nerve fibers are often centrally situated in the neurofibroma. There is no sex predilection. Korf BR. Neurofibromatosis: Phenotype, natural history and pathogenesis. The histopathological sections showed an ill-defined lesion present diffusely in the dermis and subcutaneous tissue, enveloping the normal structures, consistent with a plexiform neurofibroma. The term neurofibromatosis describes a group of genetic disorders that primarily affect the cell growth of neural tissues. NF-1 is estimated to occur in one of every 3,000 births. 1999 Mar 26;89(1):23-30. They may appear on the face, legs, or spinal cord and frequently involve the cranial and upper cervical nerves. [PubMed] [PubMed] [Google Scholar] 2. Medical, surgical, and dental histories were unremarkable. Plexiform neurofibromas. Three subtypes of neurofibromas exist: cutaneous, subcutaneous and plexiform.

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