Plexiform neurofibroma encasing vital organs

Sir,

Neurofibromatosis (NF) is one of the most common neurocutaneous syndromes characterized by varied
clinical manifestations including café-au-lait macules and nerve sheath tumors. A century ago, von Recklinghausen described neurofibromatosis in his patients—Marie Kientz and Michael Bar.[1] Diagnosis of NF is based on the criteria established by NIH (National Institutes of Health Consensus Conference), which classified neurofibromatosis into eight subgroups.[2] Four types of neurofibromas have been described, namely, cutaneous neurofibromas, subcutaneous, nodular and diffuse plexiform neurofibroma.[3] Plexiform neurofibroma is an uncommon type of neurofibromatosis, commonly seen along the branches of trigeminal nerve.[4] We herewith report a case of large, diffuse, plexiform neurofibroma in a child.

A four-year-old female child was brought with large hyperpigmented patch involving the trunk since birth, associated with underlying swelling on the left side of the abdomen. There was no positive family history. Cutaneous examination revealed a large café-au-lait macule with well-defined margins measuring 25 × 30 cm, involving the left side of the anterior abdominal wall extending to the back. There was a diffuse subcutaneous swelling underneath the café-au-lait macule measuring 10 × 12 cm. Swelling was firm in consistency, nontender and not compressible. Overlying skin showed hyperpigmentation and hypertrichosis, with follicular prominence. On the basis of these clinical findings, the patient was diagnosed with plexiform neurofibroma. Ultrasonography of the abdomen revealed soft tissue swelling. MRI of the spine showed multiple, soft tissue masses in the paraspinal region extending anteriorly into the retroperitoneum encasing the aorta and its branches and the pancreas, extending along the inferior aspect of lower ribs. MRI diagnosed it as neurofibroma. Biopsy taken from the subcutaneous swelling along with the skin confirmed the diagnosis of plexiform neurofibroma, which showed spindle cells in bundles consisting of nerve cells and fibroblasts surrounded by connective tissue stroma.

Plexiform neurofibroma (PFN) is considered to be a hamartoma than a typical tumor.[8] It may be nodular plexiform or diffuse plexiform. Diffuse plexiform neurofibroma accounts for about 5% of neurofibromatosis type I and is always congenital and pathognomonic of neurofibroma type I.[9] It is highly vascular and may involve all the layers of skin, adjacent fascia and deeper elements at times even replace the muscle and may erode the bone and infiltrate viscera. PFN is usually asymptomatic but can cause pain, impairment of function and disfigurement. Plexiform neurofibroma is commonly seen along the branches of trigeminal nerve. Orbital and periorbital regions of the face are the most common sites of involvement.[9] The risk of malignancy is 5% to 10%.[8] Tonsgard et al., have reported a case of plexiform neurofibroma, which involved retroperitoneum, mediastinum and paraspinous region.[9] The case under discussion is comparable with the case report by Tonsgard et al. Plexiform neurofibromas involving gastrointestinal tract is rare. These patients may present with epigastric pain, motility disorders, dyspepsia, anemia, hematemeses, intussusception, volvulus, intestinal perforation or bowel obstruction.

Surgical resection is the treatment required in this case. However, resection may not be advisable at this stage as the tumor involved vital structures and there are no pressure symptoms. The child has been under follow-up for the last two years and has no pressure symptoms. Prognosis is not good in this case as the tumor is likely to increase in size and cause pressure symptoms in future. The patient under study presented with large PFN with the involvement of vital organs. Hence, it is mandatory to suspect and investigate for internal organ involvement in all cases of PFN.
Hematohidrosis

Sir,

Cases of colored sweat (chromhidrosis) are very rare and are due to colored apocrine secretion. Pseudochromhidrosis refers to the condition in which initially colorless sweat becomes colored on the surface of the skin due to the action of chromogenic bacteria especially corynebacteria. Cases where colored sweat is produced in localized areas are extremely rare. Face is the commonest site and color produced may be black, violet, blue, brown, yellow or green. Red colored sweat is very rare and the pigment is lipofuscin. Bloody sweating is called hematohidrosis and is exceptionally uncommon. True hematohidrosis may occur in bleeding disorders.[1] We hereby report a case where bloody sweat discharged from the forehead episodically in a healthy young girl, who did not have any bleeding disorder.

A 12-year-old girl, resident of a village near Dharwad district of Karnataka, came to us on July 8, 2007, with the history of bleeding from the intact skin over the forehead for the last 2-years. The first episode started a few days after a horrifying incident, which she witnessed, in which a woman was beheaded by the villagers in her village. The bleeding occurred in episodes, once or twice a day, sometimes more frequently, especially when she is anxious. Sometimes, she did not get such episodes for two or three weeks. About 15-20 minutes before each episode, she gets a peculiar tingling sensation over the forehead and she becomes aware that in another few minutes she will experience the episode. Each episode started with frothy, watery secretion over the forehead soon followed by the bright red colored secretion. Each episode lasted for about 10-15 minutes and the patient remained perfectly alright during the post-episode period till the next episode. She also complained of similar episodes over her umbilical area. There was no history of bleeding from any other site. No history of ingestion of any anticoagulants, dyes or other drugs was obtained from her. She did not have any history of major medical or surgical illness in the past. No family member had similar complaints. She had attained menarche one year back. Menstrual cycles were regular and normal. On physical examination, she was anxious. Her general physical examination and systemic examinations did not reveal any abnormality. The skin over the forehead was normal. There were no cuts, abrasions or telangiectasiae [Figure 1]. There was no local tenderness. Blood or red colored secretion could not be extruded on manipulation. We witnessed one episode during our clinical examination. On gross examination, the secretion was bright red in color, less viscous than blood and it was not frank blood [Figure 2]. We collected the bloody secretion in a syringe and smeared it onto a glass slide and examined under microscope. There were plenty of erythrocytes. The left out secretion collected in the syringe was examined after 1 hour and it did not clot. Her psychiatric evaluation revealed severe depression in her. We investigated her. Her routine hemogram, blood counts, platelet count, bleeding time (2