



Dermatofibrosarcoma Protuberans Mimicking a Vascular Malformation on MRI: A Case Report of a Thigh Lesion with a CD34-Positive Spindle Cell Tumor

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Abstract

A 38-year-old male patient came to the hospital with a painless, reddish swelling on his right thigh measuring 6x5 cm. Before surgery, an MRI suggested it might be a vascular malformation, so the doctors decided to remove the area and take a tissue sample. The lab results showed a type of tumor made up of spindle-shaped cells that tested positive for CD34. This confirmed the diagnosis of dermatofibrosarcoma protuberans (DFSP). There was no sign of the tumor spreading deeper or to other parts of the body. After the surgery, the patient recovered without any complications. The doctors plan to keep a close watch with follow-up imaging and check the surgical margins. This case shows how DFSP can look like a vascular malformation on imaging and highlights the importance of examining tissue samples under a microscope and using special staining techniques for an accurate diagnosis

I. INTRODUCTION

Dermatofibrosarcoma protuberans is a rare type of skin cancer (only about 0.1% of all soft tissue cancers) that starts in the fibroblasts of the skin. It grows slowly and tends to spread locally in the skin and underlying tissue. It usually appears as a painless flat or raised area on the skin that may break open. It is more common on the trunk or upper limbs compared to the thigh. An MRI might show it as a

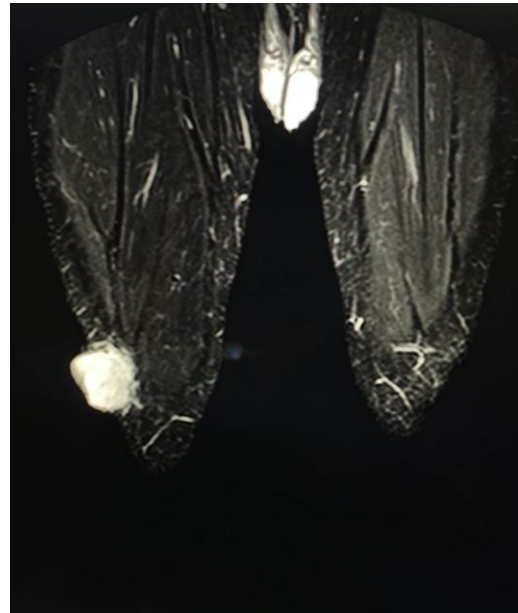
vascular-looking mass, leading to a wrong diagnosis before surgery. Early removal of the tumor with wide margins is usually the best treatment. The presence of CD34 in the tumor cells helps doctors confirm the diagnosis

II. CASE PRESENTATION

A 38-year-old man, who had no other health issues, noticed a painless red swelling on his right thigh that grew slowly over several months to 6x5 cm in size. There was no history of injury or other symptoms in the body, nor was there a family history of cancer. When examined, the doctors found a hard, non-painful lump beneath the skin without any open sores. Before the surgery, an MRI showed a bright area on T2-weighted images, which looked like a vascular malformation, with some small blood vessels surrounding the tumor. The doctors removed the tumor along with a margin of healthy tissue, and sent the tissue to the lab for testing. Under the microscope, the tissue showed a pattern of spindle-shaped cells spreading in the skin and deeper layers. The tissue tested positive for CD34 in all the cells but negative for S100, SMA, and desmin, which confirmed the diagnosis as DFSP (classic type). The surgical margins were clear, more than 2 cm away from the tumor. A computed tomography scan of the chest and abdomen didn't show any signs of the cancer spreading elsewhere in the body.



(a)



(b)

(a) PATIENT WITH SWELLING OVER HIS RIGHT THIGH ON ANTERO-LATERAL ASPECT

(b) MRI IMAGING SHOWING WELL CIRCUMSCRIBED CIRCULAR LESION IN SUBCUTANEOUS PLANE WHICH SHOWS HYPERINTENSITY ON T2 WITH FEW SUSPICIOUS SMALL SUBCUTANEOUS FEEDING VESSELS SEEN EXTENDING INTO THE LESION PERIPHERALLY

III. DISCUSSION

Most DFSP cases are located on the surface of the skin. Doctors can assess how far the tumor has spread and whether there is any involvement of nearby lymph nodes by looking at the patient's physical examination. Classic DFSP in its early stage appears as small, firm, painless plaques on the skin with thickening of the subcutaneous layer or as flat, non-raised areas. Over time, the lesion changes into a more protruding shape and appears as red or blue patches or as smooth, purple bumps. Although DFSP is unlikely to spread to distant parts of the body, it has a high risk of coming back in the same area. The tumor is made up of bundles of spindle-shaped cells growing in a storiform pattern and shows strong and widespread CD34 staining. A genetic change called $t(17;22)(q22;q13)$ causes the formation of an abnormal protein called COL1A1-PDGFB, which plays a role in the tumor's development. The way DFSP spreads, starting with growth outward before moving inward, explains why an MRI can sometimes show it as having a vascular look, resembling a honeycomb pattern of dilated blood vessels. For a tumor located on the thigh, an MRI is recommended according to NCCN guidelines to check if it has spread into surrounding muscle or bone. Common issues after treatment include wound healing problems, such as infections, difficulty

covering the surgical site requiring skin grafts, tightening of the skin, scarring, and poor cosmetic results. Surgery to remove the tumor with clear margins is essential for a good outcome (with a 10 year survival rate of 99.1%) and to reduce the chance of the tumor returning. Some new treatment options include Mohs micrographic surgery and other treatments like radiation or imatinib for patients who can't have surgery

IV. CONCLUSION

This case highlights the challenges in diagnosing DFSP using MRI and the importance of using immunohistochemistry to confirm the diagnosis. DFSP, which is made up of spindle-shaped cells, requires complete surgical removal with clear margins to achieve the best possible outcome and prevent the tumor from returning. Some of the newer treatment approaches include Mohs surgery and adjuvant therapies like radiation or imatinib for patients who cannot undergo surgery. Patients should be monitored every 3 to 6 months to check for signs of the tumor coming back (which can happen in over 50% of cases) or spreading to other parts of the body (which can occur in 10 to 15% of cases).



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CONFLICT OF INTEREST STATEMENT

The authors have no conflict of interest to declare

ETHICAL APPROVAL

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