PILOMATRIXOMA OF THE BREAST IN A PATIENT WITH TYPE 1 MYOTONIC DYSTROPHY: SUCCESSFUL SURGICAL APPROACH

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Malherbe's calcifying epithelioma is an uncommon cutaneous tumour that originates from the matrix cells of hair follicle. It was initially described by Malherbe as a benign calcifying epithelioma. Several ultra-structural and electron-microscopic studies later demonstrated its origin from matrix cells and the term pilomatrixoma was introduced. The treatment of this tumour remains mainly surgical. Malignant cases with post-surgical recurrences have been described in literature and recurrences have been related to an incomplete surgical treatment or tumour aggressiveness. We present the case of 31-year-old female patient with pilomatrixoma of the breast, which was very similar to fibroadenoma, in terms of size and other clinical features. We successfully treated this patient surgically, and the aesthetic results were good, despite the proximity of the tumour to the areola-nipple complex. Fifteen months later, the patient is doing well, free of any clinical local recurrence.

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5-HYDROXYTRYPTAMINE AND LYME DISEASE. OPPORTUNITY FOR A NOVEL THERAPY TO REDUCE THE CEREBELLAR TREMOR?

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Lyme boreliosis is caused by the spirochete Borrelia burdorferi, which is transmitted by ticks. A 59 year-old woman developed pyrexia, strong headaches, ataxia, dysarthria and tremor of the limbs after a tick bite. She was unable to work and eat on her own. She was hospitalized three times and diagnosed with cerebellar intention tremor, cerebellar ataxia, dysarthria, bilateral horizontal gaze paralysis and a central lesion of the left facial nerve. There were no pyramidal, sensory or psychiatric disturbances. The brain MRI showed multifocal leucoencephalopathy with many hyperintense areas in both hemispheres, as well as in the left superior pedunculus cerebellaris. Diagnosis was confirmed by serologic examination. Treatment with cephtriaxone, doxycycline, methylprednisolone, cephixime and ciprofloxacine was administered without effect on the tremor, ataxia and horizontal gaze paralysis. Treatment was then administered with 5-hydroxytriptamine (5-HT) in increased doses. The result of the three-month treatment with 5-HT was a gradual diminution of the tremor and the ataxia and an increase in the ability to eat, walk and work independently.

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ISOLATED CMV INFECTION CAUSING PERIANAL AND SACRAL ULCERATION IN A PATIENT WITH AIDS

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Cytomegalovirus (CMV) is a DNA virus estimated to infect 70-90% of the world's population, producing minimal symptoms in immunocompetent hosts. In the immunocompromised host, CMV infection can be potentially fatal, producing systemic or localized forms. We report the case of a 52-year-old female with acquired immunodeficiency virus (AIDS) who presented multiple sacral and perineal ulcers clinically and histopathologically consistent with CMV ulcerations. We discuss the patient's clinical presentation and histologic findings to remind physicians to consider CMV as a cause for cutaneous and systemic infection in the immunocompromised host.

Cytomegalovirus (CMV) is a DNA virus from the herpesviridae family that is estimated to infect 70-90% of the world's population (1). In the majority of immunocompetent hosts, CMV infection produces minimal symptoms (1). In the immunocompromised host, infections can be potentially fatal and manifest in either a systemic or localized form. In the localized, ulcerative form of infection, CMV is almost always found to co-infect with other herpesviridae viruses, making it difficult to establish whether CMV is involved in the pathologic formation of lesions or simply a bystander (1). Here we report a case of isolated CMV ulcerations in a patient with AIDS.

Case report

A 52-year-old female with a history of acquired

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COEXISTENT TRICHILEMMOMA AND TRICHOBLASTOMA WITHOUT ASSOCIATED NEVUS SEBACEUS

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Trichilemmoma and trichoblastoma are benign adnexal neoplasms derived from the hair follicle unit. While trichilemmomas are closely associated with the epidermis, trichoblastomas are found within the dermis and subcutaneous tissue. Both tumors have been reported to arise within nevus sebaceus of Jadassohn (NSJ). We present a 42-year-old white male with a 5 mm crusted, erythematous papule on the right occipital scalp that had been present for years. A shave biopsy was performed and read as trichilemmoma involving the biopsy base. The patient returned for follow-up 2 months later with recurrence of a crusted papule, measuring 9 mm in greatest diameter at the site of the previous biopsy. The lesion was excised for complete histologic evaluation, diagnosed as trichilemmoma with verrucoid features and associated basaloid proliferation with adnexal differentiation, again involving the biopsy base. The lesion recurred 2 months later in the form of an 8 mm multilobulated pink nodule. It was again excised and diagnosed as trichoblastoma with overlying trichilemmoma. The significance of this finding is that coexistent lesions do not necessarily necessitate a preexisting nevus sebaceous. Rather, this finding supports the notion of a common stem cell capable of differentiating toward the various portions of the hair follicle unit and adnexal structures. The idea is that any portion of the skin adnexal structure may develop out of a pluripotential germ cell and develop into a tumor.

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BIOFIBRE HAIR IMPLANT – IMPACT ON THE QUALITY OF LIFE

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Body image refers to how we feel about our bodies. It does not refer to what we actually look like, but rather to our perceptions, opinions and ways of thinking about our appearance. How we feel about our appearance is part of our body image and self-image. The hair is a significant part of this image. The problem of alopecia affects both sexes and all ages with significant sequelae. Along with androgenetic alopecia, there are forms of alopecia of various origins: traumatic, surgical, pharmacological and others. Polyamide artificial hair implant (Biofibre®) is one of the current techniques used to treat this problem.

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BIOFIBRE HAIR IMPLANT: WHAT IS NEW, WHAT IS TRUE?

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Ensuring the safety of hair implant fibers is essential. At the same time, good aesthetic quality and durability should also be considered in order to maintain expected result over the years. The main features required are biocompatibility, resistance to traction, absence of capillarity, resistance to physical-chemical stress, and low tissue trauma, in addition to good aesthetics. Biofibre® medical hair prosthetic fibers meet all the biocompatibility and safety requirements established by international standards for medical devices. They are available in 13 colors, with different lengths (15, 30 or 45 cm) and various shapes (straight, wavy, curly and afro). Biofibre® hair implants are indicated for diffuse hair loss or hair thinning in cases where an immediate aesthetic result is required, when patients request minor surgery without hospitalization, both for male and female patients, in combination with other hair restoration techniques to improve the final aesthetic result, to correct scars or scalp burns and in cases of poor donor areas. Biofibre® Hair Implant is in fact a minor surgery technique, performed under local anesthesia by either a manual implanter or an automatic machine which enables an immediate aesthetic result and the desired quantity of hair without pain or hospitalization. Clinical and histological studies have demonstrated that Biofibre® hair Implants are safe and well tolerated by patients and can be totally reversible if the need arises. This technique requires good after-care, periodical check-ups and yearly implant re-touches to maintain the best cosmetic result.

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UNCOMMON CLINICAL PRESENTATION OF KIMURA'S DISEASE AS BILATERAL RETROAURICULAR MASSES IN A YOUNG MALIAN MALE: SUCCESSFUL SURGICAL APPROACH

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We present a case of a 27 year-old Malian male referred to our hospital for two large, painless retroauricolar masses that had appeared two years earlier. Bilateral cervical painless lymphadenopathy was present at physical examination, without any other systemic symptoms. His history was relevant for bilateral Kimura's disease lesions resected 5 years earlier in the same locations. Lymphocytosis and a mild hypereosinophilia were found in routine blood tests, together with increased total IgE levels. After surgery, histology showed lymphoid infiltrates with reactive prominent germinal centres containing eosinophils, suggesting relapse of Kimura's disease, in the context of nonencapsulated fibrous proliferation with discontinuous collagen fibers, consistent with keloid. Three months after removal of retroauricular masses, abnormal laboratory findings reverted to normal. To the best our knowledge, this is the first case in literature of bilateral keloid lesions developed after surgery for Kimura Disease and harbouring its histopathologic features. Clinicians should be aware of these unusual reactive phenomena and their possible simulators.

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LANGERHANS CELL SARCOMA: AN UNUSUAL MICROSCOPIC PRESENTATION

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A 70-year-old Caucasian male presented to our clinic for a pruritic eruption progressing over several months. He complained of fatigue with a 20-pound weight loss over the past year. On presentation, the patient had browny-yellow to violaceous, purpuric, macular and papular lesions on the legs, arms, lower abdomen and back. Initial biopsy showed an angiocentric infiltrate with a suggestion of intraluminal proliferation; CD31 and Fli-1 positivity suggested either reactive angioendotheliomatosis or an unusual intravascular histiocytosis. Further excisional biopsies demonstrated perivascular collections of cells with ample cytoplasm, prominent nuclear pleomorphism and mitotic activity. The nuclei demonstrated nuclear folding, grooves and indentations. The atypical cells were S100, CD1a and CD56 positive with immunohistochemistry. A diagnosis of Langerhans cell sarcoma (LCS) was made. LCS is a rare, aggressive malignancy that can involve multiple organs including the skin, lymph nodes, lung, bone marrow, spleen, heart, and brain. The skin and lymph nodes are commonly involved, and the cutaneous presentation varies greatly. Immunohistochemistry characteristically shows CD1a and S100 positivity. CD56 expression is uncommon and often portends a poor prognosis. There is no established treatment of LCS due to its rarity. Surgery, radiation, and chemotherapy have been used with varied outcomes. Our patient was treated with prednisone with improvement of cutaneous disease. He did not develop systemic involvement, but died 1.5 years later from complications associated with heart failure. Langerhans cell sarcoma should be considered when faced with an unusual angiocentric infiltrate in which initial immunohistochemical staining results may be misleading.

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PULMONARY AND ABDOMINAL SARCOIDOSIS, THE GREAT IMITATORS ON IMAGING?

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Sarcoidosis is an insidious disorder that virtually affects every body organ. Lungs are the site most often affected (in up to 90 % of patients) followed by intra thoracic more often than peripheral lymph nodes and other sites can be involved in different percentages. The evaluation of pulmonary sarcoidosis is best performed with high-resolution computed tomography (HRCT), as traditional chest X-ray has a low resolution and can be negative or give non-significant results. Disorders such as interstitial lung diseases (ILDs), tuberculosis, lung cancer and lymphangitis carcinomatosis can manifest with similar radiological findings that can deceive clinics and radiologists. The need of a clear distinction between these conditions is important not only for diagnostic purposes but also because treatment differs significantly in different conditions. However, conventional Ultrasound (US) can be negative if small lesions are present and false negative images can result if US is not followed by a contrast-imaging technique. Contrast enhanced computed tomography (CECT) and magnetic resonance imaging (CEMRI) are preferred to detect single or multiple masses, which appear hypodense and hypointense after contrast agent administration, respectively. We think that a correct algorithm should include a thorough clinical and radiological evaluation, a definite biopsy of affected tissues revealing classical non-caseating granulomas and a certain exclusion of conditions that can give similar clinical/histophatological patterns before considering the diagnosis of sarcoidosis. Only in these cases, a diagnosis of sarcoidosis can be sufficiently achieved before starting an appropriate treatment.

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INTERSTITIAL GRANULOMATOUS DERMATITIS DEMONSTRATING SMALL, DISCRETE SKIN-COLORED PAPULES

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We report the case of a 67-year-old female with a rare variant of interstitial granulomatous dermatitis showing multiple skin-colored papules. Clinically, numerous skin-colored or reddish papules were distributed on her back and posterior thighs with itchy scaly erythema on the upper back. After topical steroid application, skin-colored papules still remained after the disappearance of itchy scaly erythema. Histopathologically, perivascular and interstitial infiltration of lymphocytes and histiocytes with occasional multinucleated giant cells were observed in the superficial and mid reticular dermis, accompanied by mild mucin deposition. Interstitial granulomatous dermatitis is similar to interstitial granuloma annulare, but can be differentiated from it by lesser degrees of collagen degeneration with mucin deposition and frequent association with arthritis or rheumatic diseases. As previously reported, multiple asymptomatic skin-colored papules are considered a rare but distinct variant of interstitial granulomatous dermatitis. Although no apparent underlying disorder has developed in the presented case, careful follow-up needs to be continued.

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MEDIUM-SIZED CONGENITAL MELANOCYTIC NEVUS OF THE FOREHEAD, GLABELLA AND TEMPLE – SURGICAL TREATMENT AND LONG-TERM FOLLOW-UP

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Congenital melanocytic nevi can be stigmatising for the patient. Larger nevi bear an increased risk for melanoma development. Large congenital melanocytic nevi may be a symptom of neurocutaneous melanosis. We report on a 5-year-old boy with an extensive hair-bearing facial congenital melanocytic nevus, covering forehead, glabella and temple region associated with unilateral brow and blepharoptosis. The lesion was excised en bloc. The resulting defect had been closed by full thickness skin graft. Healing was unremarkable and long-term follow-up over 13 years demonstrated a satisfying esthetic and functional outcome. There was no evidence of melanoma development. Surgery is an option for disfiguring larger congenital melanocytic nevi as long as esthetics and function can be preserved. Long-term follow-up is recommended due to the increased risk of melanoma.

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GIANT CONGENITAL MELANOCYTIC NEVUS IN A BULGARIAN NEWBORN

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Giant congenital melanocytic nevus (GCMN) is a rare disorder affecting 1 in 200,000–500,000 live births. Central nervous system defects such as spina bifida, meningocele, Dandy Walker malformation may accompany it and thus cause significant morbidity. Despite the related risk for malignant transformation, GCMNs may be associated with neurocutaneous melanosis, a rare syndrome in which a giant CMN or multiple smaller CMNs are accompanied by melanocytic deposition in the brain and the spinal cord. We present a case of a 5-day-old newborn with giant congenital melanocytic nevus on his back, as we discuss the diagnostic and treatment approach.

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MULTIFOCAL INFANTILE PROGRESSIVE HEMANGIOMATOSIS WITH OCULAR INVOLVEMENT: UNIQUE PRESENTATION IN A BULGARIAN NEWBORN

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Vascular disorders are considered a common finding among infants and in general, hemangioma is the most common. Diffuse neonatal hemangiomatosis is a rare and frequently fatal variant of them. We describe a case of a 2-months-old infant with multiple cutaneous hemangiomatosis and ocular involvement. To the best of our knowledge, this is the first reported case. We focus on the different treatment modalities and current diagnostic approaches.

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