Alopecia in Wegener’s granulomatosis

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Alopecia is not a distinctive clinical sign in Wegener’s granulomatosis and, as far as we know, to date no cases have been published describing this phenomenon.

CASE REPORT
We present the case of a 54 year old woman diagnosed with Wegener’s granulomatosis, who in the first stage of her disease had alopecia and improved after treatment with cyclophosphamide and prednisone.

Nine months before her admission to our service, she had had paraesthesias, and leg pain and dysfunction. Electromyography showed some signs of sensorimotor polyneuropathy. She was given prednisone for 10 days (90 mg/day) and improved partially. Five months later, she started coughing up haemoptysis sputum, and had arthralgias in both hands, constitutional symptoms, and intense and diffuse hair loss (traction positive). Her temperature was 36.5ºC, blood pressure 130/60 mm Hg, respirations 16, pulse 80 beats/min, and her weight was 44 kg.

Physical examination showed 2 cm abdomen hepatomegaly and leg distal muscular atrophy. 4/5 upper limb distal weakness, normal positional and vibratory sensitivity, paretic-spastic walk, and deep tendon reflexes increased diffusely with clonic reply. The erythrocyte sedimentation rate was 91 mm/1st h, platelets 678×10^3/l, C reactive protein 229 mg/l, rheumatoid factor 1 U/ml. A chest x ray examination showed a bilateral interstitial pattern with multiple fibre tracts of hilar origin and ulcerated segmental atelectasis. Chest computed tomography showed three small nodules located in the front segment of the right upper lobe and right middle lobe—one was 5 cm and the other two were 1 cm. We also noted scarring fibre tracts in the left upper lobe and lower lobe back basal regions. The antineutrophil cytoplasmic antibody cANCA titre was 1/160 U/ml (normal range 0–20), and antinuclear antibodies, SSA/Ro, SSB/La, RNP and Scl-70 were negative. ECA antibody cANCA titre was 1/160 U/ml (normal range 0–20), but in our patient that regimen was ineffective. Cyclophosphamide is an alkylating agent with cytotoxicity and immunosuppressive activity. Its main side effects are leucopenia, infections, vomiting and haemorrhagic cystitis. Alopecia is deemed to be one of the most common side effects of cyclophosphamide. The side effects are directly related to the doses given, so that these can be reduced with a weekly dose of a 500 mg pulse given for three months; the length of exposure to the drug may be another factor to take into account. In our patient, alopecia appeared during the active stage of the disease. Once corticosteroids and cyclophosphamide were given, we were able to control the disease activity and cranial hair loss. We believe that the pilose follicle is another organ which may be affected in Wegener’s granulomatosis by a vasculitis of the scalp vessels; and although we did not perform a scalp biopsy, it seems likely that this disease might have caused the patient’s hair loss.

The interesting aspect of this case is that the patient had Wegener’s granulomatosis and alopecia and she improved with a treatment which included prednisone and cyclophosphamide.

REFERENCES

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Paediatric Behçet’s disease in France

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Our objective was to assess the increase in the number of children with Behçet’s disease in France. To our knowledge, this survey is the most extensive reported from a single country.

Children with Behçet’s disease from any part of France were referred to one of three medical centres: Marseille, Montpellier, and Paris. Information was obtained from the medical charts and from the patient’s interview. A specific questionnaire was designed to determine the following demographic features: sex, age, city of residence, ethnicity, and familial history with complete pedigree; and the clinical variables: oral aphthous ulcers, genital ulceration, skin lesion, skin hypersensitivity plus other organ involvement—nervous system, gastrointestinal tract, eye, vessels, lungs, heart, joints, genitourinary tract, and fever. The date of onset of the disease was recorded together with the date of appearance of each symptom, and the date at which the patient met the international criteria for Behçet’s disease. A specific database was set up.

Fifty five children with Behçet’s disease met the international criteria before the age of 16 years: 33 white subjects (27 French), nine North Africans, five Turks, three West Indians, three mixed white/North African subjects, one Asian, and one Ashkenazi Jew. The male to female ratio was 0.89. The mean age of onset was 7.5 years (median 8 years, SD 4.3). The mean age at which patients met the criteria for Behçet’s disease was 11.6 years (median 12, SD 3.7; fig 1). The mean time between the appearance of the first and last criterion was 3.5 years (median 3, SD 3.7).

Initial symptoms were oral ulcers in 41 (74%) (at a mean age of 6.8 years), genital ulcers in 13 (24%) (at a mean age of 6.8 years), bipolar aphthosis in nine (16%), skin lesions in eight (14%), and uveitis in two (4%). At least two criteria were present in nine (16%) patients.

Recurrent oral ulcers were present at a mean age of 7.44 years. Genital ulceration occurred in 43 (79%) patients, at a mean age of 10.8 years. Cutaneous signs included erythema nodosum (26%), necrotic folliculitis (38%), and aphthosis (14%). Ocular signs were uveitis (36%), retinal vasculitis (24%), conjunctivitis (17%), papilloedema (7%), and keratitis (3%). Arthralgia was the main articular sign, arthritis was present in 17% of patients. Headaches were common (35%) and associated with aseptic meningitis (10%), benign intracranial hypertension (10%), and hemiparesis in two patients. Abdominal pain was reported in 40% of cases, with digestive ulceration in 14%. Ulcerative colitis was diagnosed in one patient. Venous thrombosis occurred in 21% of patients.