Case Report

Arthropathy in von Willebrand’s disease

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SUMMARY A 62-year-old woman with severe von Willebrand’s disease and a long history of joint complaints is presented. Her history, the progressive radiological findings, the demonstration of haemarthrosis and a literature review support the view that some patients with von Willebrand’s disease can suffer from an incapacitating arthropathy akin to that seen in haemophilia.

Key words: Haemarthrosis, Secondary Osteoarthrosis.

INTRODUCTION

von Willebrand’s disease (vWD) is an inheritable bleeding disorder resulting from a quantitative or qualitative defect of von Willebrand factor (vWF). This factor is a large adhesive glycoprotein that plays an essential role in the adhesion of blood platelets to the injured blood vessel wall. It also serves as a carrier and stabilising protein for coagulation factor VIII. Severe vWD is characterized by extremely low levels of vWF, low levels of factor VIII and a prolonged bleeding time.

Arthropathy in severe vWD appears to mimic that seen in haemophilia. There have been several reports (1-13) of joint problems, presumed haemarthrosis, usually of the knee, ankle or elbow. We report here a patient in whom, in addition to ankle, elbow and knee complaints, a shoulder was also affected.

CASE REPORT

A 62-year-old woman was re-evaluated in February 1987 because of joint complaints. She suffered from severe von Willebrand’s disease (bleeding time (Ivy) longer than 15 minutes, FVIII activity 2%, vWF antigen by immunoradiometric assay undetectable, ristocetin cofactor activity less than 12.5%). In several family members a milder form of the disease was diagnosed. Her parents were first cousins. In retrospect her first joint problem, a swollen right ankle, possibly occurred at the age 2. During her primary schooling (6-11 years) she had recurrent episodes of pain and swelling in the right ankle which she required about one week’s rest. She ascribed a painful left elbow, when aged 19, to a blood transfusion and has had pain and some swelling of that joint ever since. In 1982 she fell on the left knee which has continued to be intermittently painful...
and always larger than the right knee. She has noticed a deterioration in the movement of the right shoulder during the last 3 years, associated with pain on use or when lying on that shoulder. She denied early morning stiffness and had no other complaints. Full dental extraction had been undertaken without complications under transfusion cover but no other operations had been performed. She had required one admission for gastrointestinal bleeding. She bruised easily.

Physical examination revealed an obese woman with no abnormalities of the heart, lungs or abdomen. There were several small cutaneous haematomas. No tophi or nodules were present. There was an exaggerated thoracic kyphosis attributable to obesity and bad posture. The lumbar spinal movements were also restricted but without radicular pain. The right Achilles tendon reflex could not be elicited but there was no other neurological deficiency. There was no evidence of a symmetrical peripheral polyarthritis. The following joints showed a painful, limited range of movement: — right shoulder (elevation 110°, abduction 40°), elbows (20-120° right, 70-90° left); left knee (5-120° with crepitus and patello-femoral pain), and the right ankle (only 15° flexion trajec available) which was also swollen, apparently mainly by soft tissue.

In September 1987 she developed a painful effusion in the left knee after minor trauma (stepping off her bicycle); after treatment two weeks later with cryoprecipitate, joint aspiration was performed revealing old blood, very few leucocytes and no crystals. Ibuprofen provided some relief of