

Hidden Predisposition To Osteonecrosis Of The Femoral Head

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Abstract

Avascular necrosis has been documented following Disseminated Intravascular Coagulation². We are presenting such a case of bilateral avascular necrosis of femoral heads, which interestingly was followed by the spontaneous presentation of the patient's sister with a unilateral avascular necrosis of the femoral head. Both sisters presented with symptoms at the same age.

CASE REPORT

We are presenting the case of A.B., a 34 year old female who was referred to the Orthopaedic department complaining of bilateral hip pain. X-rays revealed bilateral collapse of the femoral heads consistent with avascular necrosis.

Figure 1

Figure 1: AP radiograph of the pelvis of A.B. on presentation with bilateral hip pain.



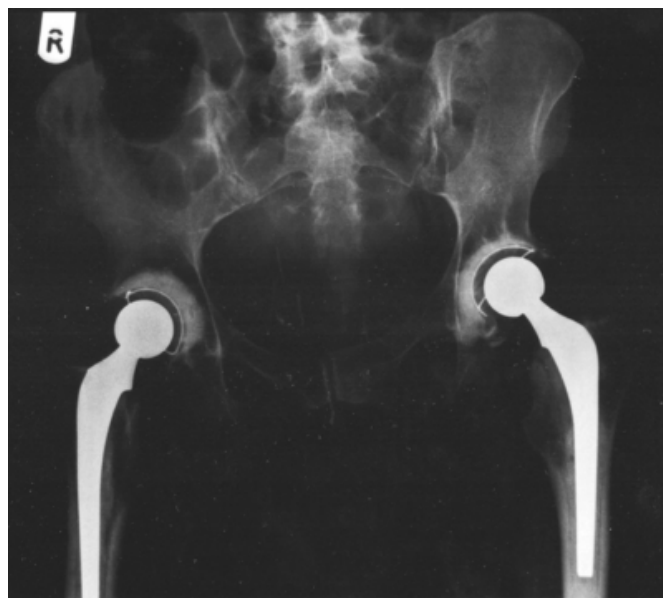
Two years previously she had given birth to her first child. The pregnancy was uncomplicated and her child was born by a normal vaginal delivery. Following delivery she was noted to have a retained placenta and suffered a massive per-vaginal haemorrhage followed by renal failure and disseminated intravascular coagulation. She was treated with supportive management on the Intensive Care Unit. Prior to

this episode she had been fit and healthy with no significant medical history.

She underwent left Charnley Elite total hip replacement 5 months later, followed by right Charnley Elite total hip replacement 10 months later. There were no complications in the peri-operative or follow-up period.

Figure 2

Figure 2: AP Radiograph of the pelvis of A.B. following bilateral hip replacement.

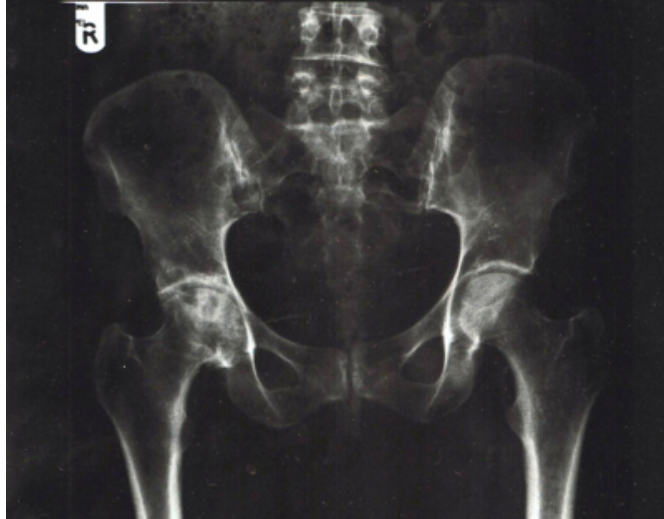


One year following A.B.'s second total hip replacement her sister F.W. was referred to the same Orthopaedic department at the age of 34 years complaining of right hip pain. She had no significant medical history and had had two pregnancies each resulting in a normal vaginal delivery with no

complications. Radiographs of the pelvis revealed collapse of the femoral head consistent with avascular necrosis.

Figure 3

Figure 3: AP Radiograph of the pelvis of F.W. on presentation with right hip pain.



Following the presentation of A.B.'s sister F.W. both sisters agreed to a full blood profile to look for any factors predisposing to Avascular Necrosis of the hip. A.B. was found to have a marginally elevated Fibrinogen level and C-Reactive protein, and a Positive Antinuclear factor (IgG 1:40). There were no significant abnormalities in the blood profile of F.W.

Figure 4

Table 1: Blood profile of A.B. and F.W. following the presentation of F.W. with hip pain.

	Normal Range	AB	FW		Normal Range	AB	FW
Hb	11.5-14.8 g/dl	13.6	12.6	Na	136-144 mmol/L	139	141
WBC	4.5-13.0 x10 ⁹ /L	5.6	5.7	K	3.6-5.2 mmol/L	4.6	4.2
Plts	169-398 10 ⁹ /L	263	322	Urea	3.0-7.0 mmol/L	5.2	3.7
RBC	3.82-4.98 x10 ¹² /L	4.54	3.93	Creatinine	60-120 umol/L	79	73
MCV	84-99 fL	93.4	99.7	Total Protein	60-80 g/L	72	68
HCT	0.36-0.46	0.424	0.392	Albumin	38-50 g/L	41	38
MCH	27.5-32.7 pg	30.00	32.10	Globulin	20-34 g/L	31	30
MCHC	30.9-34.8 pg/dl	32.10	32.20	Alanine Transaminase	0-35 U/L	13	16
Plv	1.50-1.72 cP	1.71	1.65	Alkaline Phosphatase	100-280 U/L	206	128
Neuts	2.0-7.5 x10 ⁹ /L	3.37	3.49	Gamma GT	10-50 U/L	12	11
Lymphs	1.5-3.5 x10 ⁹ /L	1.55	1.44	Total Bilirubin	0-20 umol/L	7	7
Mono	0.2-0.8 x10 ⁹ /L	0.34	0.33	Calcium	2.2-2.6 mmol/L	2.21	2.18
Eosin	0.04-0.4 x10 ⁹ /L	0.19	0.26	Adjusted Calcium	2.2-2.6 mmol/L	2.19	2.22
Baso	0.01-0.2 x10 ⁹ /L	0.06	0.05				
P.T	9.6-1.0 secs	10.6	9.6	Apolipoprotein A	0.09-2.03 g/L	1.0	1.3
APTT	24.0-30.0 secs	27.00	25.00	Apolipoprotein B	0.34-1.00 g/L	0.5	0.9
Fibrinogen	1.8-4.5 g/dl	4.79	3.25	Apolipoprotein B/A ratio			
THSA		Normal	Normal	Antinuclear Factor		Positive IgG 1:40	Negative
				Smooth Muscle		Negative	Negative
				Gastric Parietal Cell		Negative	Negative
Factor VIII	0.5-1.5 I.U./ml	1.44	1.30	Mitochondrial		Negative	Negative
DRVVT	0.8-1.1	1.1	1.0				
KCT	0.8-1.1	1.0	1.0	Pemphigus/Pemphigoid		Negative	Negative
FSL APTT	25-35 secs	34.3	27.0	Rheumatoid Factor	0-25 I.U./mL	1	1
Lupus Anticoagulant		Absent	Absent	C-Reactive Protein	0-6 mg/L	12.8	2.2
Ant-Thrombin III	0.84-1.17 I.U./ml	0.88	0.97				
Protein C (venom/Chrom)	0.71-1.42 I.U./ml	0.73	1.15	Cardiolipin	0-15 u/L	<5	<5
Protein S Ag (Total)	0.71-1.36 I.U./ml	0.88	0.79				
Protein S Ag (Free)	0.64-1.31 I.U./ml	0.76	0.82	Hep 2 ANF Pattern		Negative	
APC-R Ratio (V corrected)	2.11-2.51	2.37	2.29				
Prothrombin 20210A Allele		Absent	Absent	Cholesterol	3.5-6.5 mmol/L	2.9	5.1
Factor V Leiden		Absent	Absent	Triglycerides	0-1.8 mmol/L	0.25	0.76
APCR		3.21	2.82	TSH	0.3-5.0 uU/mL	0.92	0.81

DISCUSSION

Avascular necrosis within the skeletal system has been associated with pregnancy₁ and reported following episodes of Disseminated Intravascular Coagulation (DIC)₂ although it is most commonly attributed to trauma, alcohol intake and the use of corticosteroids₃. In the Case of A.B. DIC secondary to a massive haemorrhage was the presumed causative factor involved in the bilateral collapse seen in the femoral heads, however following the presentation of A.B.'s sister with similar symptoms of hip pain and radiographic appearances the possibility of a genetic predisposition within the family was raised, although no abnormalities in the blood profile were identified in both sisters. Thrombophilia and hypofibrinolysis have been suggested as genetic causes of osteonecrosis in both adults and children₄. These disorders pre-dispose to the formation of fibrinous clots which obstruct venous drainage of the bone and cause venous hypertension. Cell death within the bone results from the ensuing ischaemia. Glueck et al (1997) state that most cases of osteonecrosis thought initially to be idiopathic will prove to be caused by a thrombophilic or hypofibrinolytic condition on further investigation₄. One marker of this is the

Mutant factor V Leiden gene. Freiberg et al advocate screening for resistance to activated protein, protein C deficiency, protein S deficiency, anticardiolipin antibodies IgG and IgM and Lupus anticoagulant⁵. They recommend testing for the mutant Factor V Leiden gene in those with abnormal resistance to Protein C. In those patients in whom the disease is picked up early they report retardation or reversal of both the patients symptoms and underlying osteonecrosis with early medical intervention with androgenic steroids or anti-coagulant therapy⁶. In our patients only A.B was found to have a positive IgG ratio of 1:40, with marginally elevated fibrinogen and C-reactive protein levels, however all other investigations were normal.

The Apolipoprotein B/Apolipoprotein A1 ratio indicates cholesterol transport and has been used as a marker of coronary heart disease. It has been suggested by Miyanishi et al (1999) as a predictor of risk of non-traumatic AVN. In A.B. and F.W. these ratios were 0.5 and 0.69 respectively⁷. In comparison with Miyanishi's results which found an apo B/apo A1 ration of 0.85 and 0.63 for Non-traumatic AVN and Osteoarthritis respectively it does not appear that either sister would have been predicted to be at increased risk of AVN using this method.

Most of the familial, genetic and pathological causes have been excluded in these cases, leaving no known cause for the presentation of both sisters with avascular necrosis of the femoral head. We believe there is some unknown genetic or familial predisposition which can present in members of the same family at a particular age, in this case at the age of thirty four years.

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