CARDIOCUTANEOUS SYNDROME PRESENTING AS A HEMIPARESIS

S.E. Baron, J. Britton, M. Saleem, S. Clark, R. Sheehan-Dare. Dermatology and Cardiothoracic Surgery Department, The General Infirmary at Leeds, Leeds, LS1 3EX

Cutaneous pigmented lesions may be associated with cardiac abnormalities in the LEOPARD, NAME, LAMB and CARNEY syndromes. These syndromes have common features including cutaneous lentigines, which develop in infancy or early childhood and cardiac abnormalities, most frequently atrial myxomas and conduction defects, which develop in the teenage years.

We describe a case of a child who initially presented to the dermatologists with multiple facial lentigines. He later presented aged 20 years to the Accident and Emergency department following an episode of collapse and confusion. Shortly after arrival he developed right-sided focal seizures and neurological examination revealed marked right-sided weakness and an up-going right plantar reflex. On examination he had multiple facial pigmented macules with extension onto the trunk and a subcutaneous nodule on his left shoulder.

On investigation his ECG showed right bundle branch block, left axis deviation and left ventricular hypertrophy. A head CT scan revealed an extensive infarct in the left middle cerebral artery territory. An urgent echocardiogram showed a large left atrial tumour prolapsing across the mitral valve almost obstructing blood flow. The tumour was resected and confirmed histologically to be an atrial myxoma. The cutaneous nodule on the left shoulder and two of the pigmented macules were later excised and these were histologically a cutaneous myxoma and lentigines.

Our case is unfortunately the second of cutaneous lentiginosis, atrial myxoma and cutaneous myxoma to present as a hemiparesis. It is therefore important for dermatologists and paediatricians to recognise the characteristic facial lentiginoses that develop in early childhood. The atrial myxomas tend to develop in later childhood so it is imperative for these children to have annual cardiac evaluation to reduce morbidity and mortality.