



Abstract 524 Figure 2a



Abstract 524 Figure 2b

Evaluation for thrombophilia and tests for specific predisposing conditions were normal. The patient was treated by third generation cephalosporin, clindamycin and low-molecular weight heparin. Two months after the initial diagnosis, the evidence of a small focal thalamic hemorrhage was noted on MRI. Recanalization of venous occlusions were detected on MRI venography. Low-molecular weight heparin therapy was changed with oral acetylsalicylic acid.

**Conclusion** Since CSVT is an underrecognized cause of intracranial pathologic abnormality in the pediatric population, we wish to draw the attention of pediatricians to the importance of a high level

of suspicion for this diagnosis and the preferred diagnostic method which is MRI with venography.

#### 525 PAEDIATRIC MOYAMOYA SYNDROME ASSOCIATED WITH IRON DEFICIENCY ANAEMIA: A CASE REPORT AND REVIEW OF THE LITERATURE

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**Introduction** Moyamoya syndrome is a rare, progressive disorder characterised by stenosis of the vasculature of the Circle of Willis with formation of a tortuous collateral system. The aetiology may be idiopathic, or as a consequence of a number of associated diseases. No previous cases secondary to iron-deficiency anaemia have been reported.

**Aim** To report a case of Moyamoya syndrome secondary to iron-deficiency anaemia, and review the literature with regard to the haematological associations of Moyamoya syndrome.

**Method** The relevant case notes were reviewed. Key databases were searched for studies including the target phrases "Moyamoya syndrome" and "anaemia".

**Results** *Case history:* A 3 year old girl presents with a 12 hour history of right upper limb weakness and slurred speech. A background of pallor, daily vomiting and abdominal pain is elicited. Reduced power in the right upper and lower limbs and a right-sided upgoing plantar are noted on examination. Investigations reveal a haemoglobin of 1.9g/dL, with an iron-deficiency pattern. A faecal occult blood test is positive. Magnetic Resonance (MR) imaging shows focal middle cerebral artery territory ischaemia. MR Angiography shows Moyamoya vessels. Extensive investigations for associated pathologies are normal.

*Literature review:* Sickle cell disease is the most common haematological cause of the Moyamoya syndrome, whilst thalassemia, Fanconi's anaemia and paroxysmal nocturnal haemoglobinuria are also reported. The likely pathogenesis in these instances is intimal proliferation in response to impaired arterial flow.

**Conclusion** We propose that severe iron-deficiency anaemia may result in Moyamoya syndrome through the mechanism of disrupted intracerebral arterial flow.

#### 526 PROTOCOLS FOR THE TREATMENT OF HODGKIN LYMPHOMA - CASE REPORT

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**Introduction** Hodgkin lymphoma is malignant disease of cells in the lymphatic system that is characterized by proliferation of Reed - Sternberg cells. Symptoms include the painless swelling of lymph nodes, spleen, or other immune tissue. Other symptoms include fever, weight loss, fatigue, or night sweats. Also called Hodgkin disease.

**Objective** Was the presentation a case with Hodgkin lymphoma, treated in Pediatric Clinic in Prishtina at the Hemato-Oncology Unit.

**Methods** Patient, male child, 9 years old, came in the clinic because of painless enlargement of lymph nodes on the left side of the neck. Other examinations, according to systems have been in normal range. Diagnosis is made based on anamnesis, clinical examination, laboratory, ultrasound, CT, histopathology with immunochemistry, and bone marrow aspiration. The surgery has been made and according to path-histological and immunochemistry analyses the

diseases has been diagnosed as Type - Classic of Hodgkin lymphoma (mixed cellularity).

**Results** The treatment is done according to protocol for Hodgkin lymphoma, with three cycles of OEPA (Prednisone, Vincristine, Doxorubicine, Etoposide), and three cycles of COPP (Prednisone, Procarbazine, Vincristine, Cyclophosphamide). After therapy, the PET - scan has been made and its results show that nowhere in the body is present malignant infiltration.

**Conclusion** OEPA and COPP are very efficient therapy in the treatment of Hodgkin lymphoma.

## 527 CHEMOTHERAPEUTIC TREATMENT OF RHABDOMYOSARCOMA BOTRYOIDE

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**Introduction** Rhabdomyosarcoma (RMS) is malignant tumor which arises from embryonic muscle cells. RMS is the most common type of soft-tissue cancer in children. It can develop on children at any age, but it's most common in those between 2 and 6 years old and 15 and 19 years old.

**Purpose** Presentation of the case with RMS botryoides, which was presented to the Hematology/Oncology Unit at Pediatric Clinic and has received chemotherapy treatment.

**Materials and Methods** A female child, 2 years old, who was initially admitted in Clinic University Centre of Tirana, with presence the mass of tumor in vulvovaginal region. The tumor mass appeared after the birth and it has been increasing in size since then. At University Clinic Center of Tirana the patient has been operated, and the tumor has been removed from her. Detailed Histopathological analyses has shown that the removal part of her was botryoides Rhabdomyosarcoma. The diagnose of the patient was made based on anamnesis, clinical examination, laboratory tests, radiological images, bone marrow aspiration, histopathological analysis. After the diagnoses it was indicated the treatment protocol of chemotherapy for extracranial rhabdomyosarcoma VAC.

**Conclusion** The total Resection of the tumor mass, the application of the protocol for RMS botryoides, supportive care, monitoring of the chemotherapy toxicity has resulted in absence of minimal residual disease, which is confirmed by the follow up of her clinical status, laboratory tests, radiology tests, that has resulted the absence of secondary deposits.

## 528 IATROGENIC TRACHEAL RUPTURE IN A CHILD: CASE STUDY AND REVIEW OF LITERATURE

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Tracheal rupture is rare in childhood and optimal treatment is not clear. A 14-year old boy was admitted to a local hospital after sudden loss of consciousness. He underwent endotracheal intubation and was referred to our hospital. The patient's history revealed that he had voluntarily inhaled butane gas. The physical examination was consistent with coma and cardiogenic shock, and the chest x-ray showed pulmonary edema. The patient was admitted to the intensive care unit, and diuretic and inotropic therapy was started. In the third hour of monitoring of the patient under mechanical ventilation, subcutaneous emphysema and pneumothorax at the

right hemithorax were observed without deterioration of the vital functions. Thoracic computed tomography (CT) scan findings were consistent with tracheal rupture. The patient was monitored conservatively without surgery. On the fifth day of hospitalization, his tube was removed, and he was discharged on the twelfth day with a positive prognosis. In this study, a tracheal rupture case after endotracheal intubation is presented in which the patient recovered completely with conservative therapy.

## 529 OSTEOPENIA IMPERFECTA IN A DEFINED BIRTH COHORT: REGIONAL EXPERIENCE AND THE NEED FOR A NATIONAL PLAN

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**Background and Aims** We aim to analyse the clinical characteristics of OI within a 10 year regional birth cohort in Ireland and to highlight the management challenges and the need for a National level referral and treatment structure.

**Methods** Case notes of all patients with OI from 2002 to 2011 were reviewed for clinical characteristics, radiological findings, results of molecular genetics and the comprehensive management including treatment received abroad.

### Results

Case1: Female type III OI born by caesarean section at term noticed to have several fractures in-utero and infancy; now 10 years with short stature & respiratory problems.

Case2: Male type IV OI, born by CS at term. Multiple antenatal fractures noted on postnatal skeletal survey. Presently 6 years old.

Case3: Male type III OI, presented at birth with multiple fractures, and significant ligamentous laxity. Presently 4 years old.

Case4: Female type V OI, born by CS at term. Diagnosed at 9 weeks when presented with fractures. Developed subsequent crush fractures of vertebrae despite being on pamidronate infusions; presently 3 years old.

Case5: Female type IV OI, born by CS at term with antenatal suspicion of skeletal dysplasia. Presently 2 years old with associated developmental delay.

Case6: Male type IV OI, followed up since 27 weeks gestation and intrauterine death following the development of non-immune hydrops at 34 wks.

**Conclusion** As more children are surviving nationally there is a need for a single specialised and comprehensive paediatric metabolic bone diseases unit providing a structured quality of care and avoiding travel abroad.

## 530 ACQUIRED MICROCEPHALY IN A CHILD WITH ABSENT SPEECH: HOW TO STUDY?

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**Introduction** Microcephaly is defined as reduce of the head circumference less than 2 SDS for age, sex and gestation.

**Purpose** Three different cases of acquired microcephaly and absent speech are presented with the purpose to find the better form to study patients with this clinical association.

### Results

Case 1 - Eight-year old girl, has acquired microcephaly since 15 months of age, absent speech, delay motor skills, seizures and angelman-like features. Chromosome 15 study confirmed Angelman Syndrome.