

joint and visceral involvement. Usually the skin is not thickened and there is multiple large nodules and tumours. Clinical features appear after one year and slowly progress. There is usually no diarrhea, chest infection and growth retardation. Survival is usually prolonged beyond 30 years.

No specific treatment is available for ISH. Early surgical excision is recommended by some authors for those lesions that either present a significant cosmetic problem or produce some degree of functional impairment. However, excision may be followed by recurrences. Spontaneous regression has been reported in some cases, but long-term regression is unlikely and tumors continue to increase in size and number. Intralesional steroid injection may reduce the size of early lesions. Capsulotomy of joints may show some temporary, beneficial effect. Gingival overgrowth may be treated with partial gingivectomy. Oral D-penicillamine has been used in some cases with apparent improvement in joint mobility and flexibility. Therapeutic trials with dimethyl sulfoxide, ketotifen and calcitriol have been given in individual cases. Genetic counselling should be done as there is 25% chance of development of disease in future offsprings.

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Fenestrated Angiocatheter for Extensive Subcutaneous Emphysema

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A minimally invasive technique using fenestrated angiocatheters has been described in adult literature for the treatment of extensive subcutaneous emphysema. Here we report a 30 month old infant who developed extensive subcutaneous emphysema and pneumomediastinum, which was relieved by application of microdrainage catheters.

Key words: *Acute respiratory distress syndrome, Pneumomediastinum, Subcutaneous emphysema.*

Subcutaneous emphysema is a known complication of invasive procedures, some surgical interventions, and medical conditions with airway obstruction. It may also occur during

mechanical ventilation for acute respiratory distress syndrome (ARDS) and can be traced to pneumomediastinum with or without a pneumothorax. Any condition that creates a gradient between intra-alveolar and

perivascular interstitial pressures can create pneumomediastinum with subcutaneous emphysema. The condition causes disfigurement, discomfort and anxiety, but rarely airway compromise and respiratory failure. The management is generally conservative but for severe cases, micro-drainage is described in adult patients.

CASE REPORT

A 30-month old boy with acute lymphoblastic leukemia, who developed febrile neutropenia and severe respiratory failure requiring intubation after the first induction chemotherapy. Despite broad coverage antibiotic, antifungal and antiviral therapy, the child developed ARDS. Open lung and permissive hypercapnia strategy was followed. Despite the lung protective strategy he developed a left sided pneumothorax which was easily relieved by a pigtail catheter, followed by the development of pneumomediastinum and massive subcutaneous emphysema from the head towards the scrotum. Escalated ventilatory requirements without any other identifiable cause suggested that his massive subcutaneous emphysema combined with pneumomediastinum was creating restrictive pattern for ventilation. Literature search revealed a simple potential solution with modified-fenestrated angiocatheters. We chose to use 18 gauge catheters due to the size and skin condition of the child. Two angiocatheters were prepared at the bedside under sterile conditions as described by Beck, *et al.* [1], with minor modifications. Due to the smaller size of the chosen catheter, only five holes were opened using a scalpel, three at the top and two at the bottom of the plastic while the needle was in situ. The area of insertion was also modified. Instead of using the infraclavicular region we chose to stay away from the vascular port site and decided to use the area of most swelling which corresponded to low thoracic region at the midaxillary line. After preparation of the area with chlorhexidine scrub and draping, the modified catheters were inserted subcutaneously with 40-45 degree angle, about 0.5-1 cm aiming cephalad, and redirected in parallel plane to full insertion. The angiocatheters were taped to skin with a clear adhesive tape and connected to underwater seal drains aiming for a closed system and for allowing direct visualization of air evacuation. Compressive massage was applied by nurses towards the draining angiocatheters every four hours. Drainage of air was enhanced with massaging, which was verified by the simultaneous observation of bubbling in the water seal chamber. The angiocatheters were kept in place for 24-48 hours until the cessation of bubbling. Within 24 hours, substantial improvement of ESE was observed. The pneumomediastinum also decreased, albeit more slowly, with complete resolution occurring in five days. The ventilator support could be weaned, and the patient was

successfully extubated in five days. There were no associated complications with insertion of the subcutaneous angiocatheters. The patient was transferred back to the wards one week after the extubation. He remains well at home on his maintenance chemotherapy regimen six months after his intensive care stay.

DISCUSSION

Various invasive and uncomfortable techniques have been used to treat extensive subcutaneous emphysema, with potential of inducing subcutaneous emphysema themselves. Infraclavicular incisions, placement of additional chest tubes through either intrapleural or subcutaneous route, and insertion of large bore subcutaneous drains with or without suction, and tracheostomy were traditional options for treatment. The successful micro-drainage of the subcutaneous emphysema with simply constructed angiocatheters was first described by Beck, *et al.* [1]. This was followed by three other adult case reports [2-4]. Leo, *et al.* [5], also reported their experience with microdrainage catheters in their retrospective review of 12 patients from the European Institute of Oncology database after major thoracic surgeries. The procedure was reported to be effective and free of complications [5].

Our patient required increasing ventilatory pressures despite tolerance of permissive hypercapnia after development of ESE and pneumomediastinum, and only after microdrainage we were able to wean the supplemental oxygen and ventilatory pressures. This observation suggests that subcutaneous air can potentially create a restrictive defect during ventilation of already diseased lungs. We observed that this minimally invasive technique was simple, and effective in our patient, and does not have a potential for serious complications. We propose that this procedure may be considered as a first line therapy for symptomatic ESE in sick children.

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Disseminated Rhabdomyosarcoma Presenting as Hypercalcemia

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Hypercalcemia may be seen in children with malignancy. We report a 4-year old child with rhabdomyosarcoma that initially presented with signs of hypercalcemia and bone involvement.

Key words: Hypercalcemia, Metastasis, Rhabdomyosarcoma.

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Hypercalcemia is one of the severe complications of malignant diseases. For patients with rhabdomyosarcoma particularly with bone metastasis, hypercalcemia may be an initial sign [1]. We report a child with rhabdomyosarcoma that initially presented with signs of hypercalcemia and bone involvement, but the primary mass could not be detected.

CASE REPORT

A four-year old girl presented with complaints of high fever and difficulty in walking. The patient had generalized pain increasing with movement in her extremities and experienced fatigue and weakness. Laboratory tests showed hypercalcemia (15.3 mg/dL) increased LDH (1261 UI/l) and anemia (8 g/dL). Platelet and white blood cell count were normal. Routine serum biochemistry, including parathormone levels (14 pg/mL, normal 15-68 pg/mL) were normal. In addition, 24-hour-urinary calcium excretion (117 mg/24h, normal 80-320) and 24-hour-urinary VMA/creatinin ratio (8.4 mg/g, normal <13) were also normal. Peripheral smear and bone-marrow did not reveal any atypical or blast cells.

Skeletal radiographs revealed generalized osteopenia, collapse of several vertebral bodies, and multiple lytic lesions in long bones with permeative destruction. Abdominal USG was normal. CT scans of the thorax and pelvis also revealed lytic lesions. ^{99m}Tc whole body bone scintigraphy revealed multiple foci

increased or decreased uptake of radioactivity. Iliac bone biopsy revealed some massy, pleomorphic tumoral tissue infiltration composed of tumoral cells with hyperchromatic nuclei, some of which were clear and large, some fusiformly extended, and some with large eosinophilic cytoplasm stemming from atypical cell bundles resembling sporadic rhabdomyoblasts and with a sporadic storiform pattern. An immunohistochemical study showed diffuse staining with vimentin and positive staining with MyoD1 and desmin, only in rhabdomyoblasts, therefore resulting in the histopathologic diagnosis of rhabdomyosarcoma.

Intravenous hydration, furosemide at a dose of 2 mg/kg/day, pamidronate 1 mg/kg/dose twice a week (with a cumulative dose of 4 mg/kg) and calcitonin nasal spray 200 IU twice a day were given for hypercalcemia. No side effects were seen. Serum calcium level normalized (9 mg/dL) on the 14th day of treatment, and treatment for hypercalcemia was stopped. Chemotherapy was given to the patient with a modified EVAC regimen. However, despite initial improvement, the disease progressed to leptomeningeal metastasis. The patient died of progressive illness, having intractable convulsions and worsening consciousness.

DISCUSSION

Up to one-third of adult patients with cancer may develop hypercalcemia during the course of their disease [2]. The incidence of hypercalcemia in rhabdomyosarcoma varies