CASE REPORT

Superior vena cava syndrome in children

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Abstract Superior vena cava syndrome (SVCS) is rare in childhood. 18 cases of SVCS were seen in children ranging from 3–14 years with a mean age of 8.8 years. There were 15 males and 3 female children. Diagnosis could be confirmed in 17 cases as one child succumbed to severe respiratory distress without a definitive diagnosis. The commonest cause of SVCS was lymphoma. Non-Hodgkin's lymphoma (NHL) was more common than Hodgkin's disease. In two cases the final diagnosis was tuberculosis of mediastinal lymph nodes. The diagnosis was confirmed by cervical lymph node biopsy in 6 cases, mediastinal biopsy in 6 cases and bone marrow aspiration in the remaining 5 cases. Intravenous Dexamethasone provided relief of symptoms in 13 patients. None of the children received emergency radiotherapy. Anti-tubercular treatment produced complete cure in the two patients with tubercular mediastinal lymphadenopathy.

Keywords Superior vena cava syndrome · Superior mediastinal syndrome · Lymphoma · Tuberculosis

Introduction

Superior vena cava syndrome (SVCS) refers to the signs and symptoms of compression of superior vena cava. Concomitant obstruction of airway leading to stridor is termed as superior mediastinal syndrome [1]. Superior vena cava is the major vessel for drainage of venous blood from head, neck, upper extremities and upper thorax. It is thin walled and compliant and therefore vulnerable to compression by any space occupying lesion in the vicinity. The clinical features of superior vena cava compression include swelling of the face, neck and upper torso, prominence of the neck and superficial chest veins, cyanosis or plethora, stridor, dyspnoea, cough, chest pain and headache. SVCS is rare and especially more so in children. From its earliest description by William Hunter in 1757, 502 cases were reported from 1757 to 1949, of which, only 4 were in children [2, 3]. The cause of obstruction also varies according to the age of the patient. In children, surgery for congenital heart disease accounts for majority of cases and mediastinal tumors are rare. There are very few reports in Indian literature on SVCS in children. In this report we share our experience with 18 cases of SVCS in children.

Material and methods

This prospective study was carried out on the children admitted with a diagnosis of superior vena cava syndrome in the Department of Pediatrics, Institute of Medical Sciences, Banaras Hindu University, from January 2004 to December 2006. The presenting signs and symptoms along with laboratory investigations, diagnostic procedures and type of treatment given were noted in all the cases. Baseline

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hematological investigations and radiological evaluation including chest radiograph, thoracic computed tomography and echocardiography were done as required. Diagnostic procedures included fine needle aspiration cytology (FNAC), lymph node biopsy, mediastinal biopsy, bone marrow aspiration/biopsy and thoracocentesis. The patients were followed up in the pediatric hematology-oncology clinic.

Observations

An 18 children, of whom three were females, were admitted with a diagnosis of superior vena cava syndrome. Mean age of the patients was 8.8 years (range 3–14 years). Facial swelling and prominent superficial chest veins were major presenting symptoms being present in all the 18 cases (100%) while cough was present in 16 cases (88.9%). Dyspnoea and stridor were present in half to two third of the patients and headache was present in one third of the cases (Table 1). The longest mean duration of presenting symptoms was 90 days for cough and prominent chest veins, whereas, it was shortest (15 days) for dyspnoea and stridor. Pleural effusion was seen in 7 cases and pericardial effusion in 2 cases. All the cases had mediastinal widening. In 17 patients a definitive tissue diagnosis could be obtained. Diagnosis included non Hodgkin's lymphoma (NHL) in 8, Hodgkin's disease (HD) in 2 (nodular sclerosis and mixed cellularity), acute lymphoblastic leukemia (T- cell ALL) in 3 and acute myeloid leukemia (M 2 type) and round cell tumor in one each. 2 cases aged 8 and 14 years respectively who presented with facial swelling, prominent chest veins and cough were provisionally diagnosed as lymphoma. However, mediastinal biopsy revealed a diagnosis of tuberculosis in both the cases. One patient with a suspected diagnosis of lymphoma, succumbed to his illness before a definitive diagnosis could be made. FNAC and biopsy of cervical/axillary lymph node could establish the diagnosis in 6 cases. Mediastinal biopsy was required in further 6 cases and bone marrow examination established the diagnosis in the remaining 5 cases. Of the 18 cases, 13 received

intravenous corticosteroids at admission or before undergoing diagnostic procedures to relieve the symptoms. Intravenous dexamethasone 0.6 mg/kg was able to relieve stridor, dyspnoea and reduce facial puffiness within 24 hours. The two cases who were subsequently diagnosed as tuberculous lymphadenopathy did not require intravenous dexamethasone as there was no dyspnoea or stridor. Patients of NHL and Hodgkin's disease received 6 cycles of CHOP (cyclophosphamide, adriamycin, vincristine, prednisolone) and COPP/ABVD (cyclophosphamide, vincristine, procarbazine, prednisolone/ adriamycin, bleomycin, vincristine, dacarbazine) respectively. All the three patients of T- cell ALL received the modified UK MRC X protocol for high risk patients. Two patients, one each of AML (M2 type) and germ cell tumor refused treatment due to financial reasons and were lost to follow up. Those with tuberculosis received anti tubercular treatment and showed complete response. Their symptoms gradually improved over a period of two weeks. One patient with a provisional diagnosis of non hodgkin's lymphoma could receive only one cycle of chemotherapy and expired due to severe respiratory distress. The cause of the respiratory distress was compression of the airway due to cervical and mediastinal lymphadenopathy. None of the patients received emergency radiotherapy.

Discussion

Compression of structures in the superior mediastinum is referred to as the "superior mediastinal syndrome" and is almost synonymous with the superior vena cava syndrome in which the venous compression is stressed [4]. The condition is rare in children and adolescents but is a medical emergency requiring urgent treatment [5]. Early in its course, caval compression may be asymptomatic and minimal signs overlooked or disregarded. However, complete venous blockage may develop suddenly leading to catastrophic event. In the present series also the patients presented earlier to the hospital when dyspnoea and stridor were present. Minor symptoms like cough and prominent

Table 1 Symptoms at initial presentation in patients with SVCS

Presenting symptoms	No. of patients (%)	Mean duration in days (Range)
Dyspnoea	12 (66.7)	15 (7–20)
Cough	16 (88.9)	90 (30–120)
Facial swelling	18 (100)	60 (30–90)
Prominent superficial chest veins	18 (100)	90 (30–120)
Stridor	10 (55.6)	15 (7–20)
Chest pain	8 (44.4)	30 (20–45)
Headache	6 (33.3)	20 (15–45)

superficial chest veins were disregarded for a longer time. Two patients presented very late in the course of their illness with fatal outcome.

In the present study the most common cause of superior vena cava syndrome was lymphoma. Non Hodgkin's lymphoma contributed more cases than Hodgkin's disease. In one Indian study [6] all the cases had a malignant etiology. T cell ALL was the most common cause followed by lymphoma. Similar findings were reported in the series by Yellin et al [7] where T cell ALL was again a major contributor. In a series of 175 children and adolescents with SVCS [2], mediastinal tumors were the commonest cause of primary non iatrogenic SVCS. Iatrogenic causes included cardio vascular surgery, ventriculoatrial shunting for hydrocephalus and catheterization of the superior vena cava. Issa et al [8] reported 10 cases with SVCS of which 8 had lymphoma. In the present series, two patients had tubercular mediastinal lymphadenitis. Tuberculosis is a very rare cause of SVCS now days. However, it has been reported in older studies. The benign causes listed in previous reports include goiter, tuberculosis, syphilis and idiopathic mediastinal fibrosis [9]. Benign causes have declined now because of early detection and treatment of infectious mediastinal lesions.

Since SVCS is an emergency condition, over-enthusiastic attempts to establish a diagnosis can be life threatening. Establishing a working diagnosis is highly desirable [10]. Physical examination, plain radiography, bone marrow studies and biopsy of a peripheral node under local anaesthesia are simple and safe procedures. If the results of these tests are non contributory and the clinical state of the patient is critical, the treatment must not be delayed [8]. Thoracotomy and mediastinal biopsy are fraught with danger because of serious anaesthetic risks and risk of massive hemorrhage from injury to dilated collateral veins [11]. In contrast, venography was found to be safe and not to have any major complications [12].

Rapid high dose irradiation has been shown to provide symptomatic relief in 24–72 hours without producing "radiation edema" [13]. Some workers have also used simultaneous radiation and chemotherapy to ensure rapid regression of the tumor. In cases where SVCS is caused by highly chemotherapy sensitive tumors like lymphoma and leukemia as is the case in pediatric patients, chemotherapy may be as effective as radiation [6]. Radiation may increase the toxicity in such cases. In a study by Nitschke et al [14] a combination of prednisolone with vincristine was found to be fast and effective in NHL. The simultaneous use of corticosteroids has an additional advantage because of their anti inflammatory effect. In the present study also dexamethasone was administered to 13 of the 16 patients of SVCS with malignant etiology with good response. Although it

was given before biopsy it did not cause any difficulty in interpretation of the histology. Radiotherapy was not used in any patient because of unavailability during emergency. Recently, percutaneous transluminal angioplasty has been used to maintain the patency of superior vena cava in cases of obstruction by malignant and benign causes [15]. If other treatment modalities have been exhausted, surgical placement of bypass grafts can be tried.

References

1. Lange B, O'Neill JA, Goldwin JW, Parker RJ, Ross AJ (1997) In: Piazzo PA, Poplack DG, eds. Principles and Practice of Pediatric Oncology. Philadelphia; Lippincott-Raven, 1025–1049
2. Janin Y, Becker J, Wise L, Schneider K, Schwartz D, So H (1982) Superior vena cava syndrome in childhood and adolescence. A review of literature and report of three cases. *J Pediatr Surg* 17:290–295
3. McIntire FT, Sykes EM Jr (1949) Obstruction of superior vena cava. A review of literature and report of two personal cases. *Ann Intern Med* 30:925–955
4. D'angio GJ, Mitos A, Evans AE (1965) The superior mediastinal syndrome in children with cancer. *Am J Roentgenol Radiol Therap Nuclear Med* 93:537–544
5. Piastra M, Ruggiero A, Caresta E, Chiaretti A, Pulitano S, Polidori G, Riccardi R (2005) Life threatening presentation of mediastinal neoplasms: report on 7 consecutive pediatric patients. *Am J Emerg Med* 23:76–82
6. Arya LS, Narain S, Tomar S, Thavaraj V, Dawar R, Bhargava M (2002) Superior vena cava syndrome. *Indian J Pediatr* 69: 293–297
7. Yellin A, Mandel M, Rechavi G (1992) Superior vena cava syndrome associated with lymphoma. *Am J Dis Child* 146: 1060–1063
8. Issa PY, Brihi ER, Slim MS (1983) Superior vena cava syndrome in childhood: report of ten cases and review of literature. *Pediatrics* 71:337–341
9. Chen JC, Bongard F, Klein SR (1990) A contemporary perspective on superior vena cava syndrome. *Am J Surg* 160: 207–211
10. Lokich JJ, Goodman R (1975) Superior vena cava syndrome: clinical management. *JAMA* 231:58–61
11. Northrip DR, Bohman BK, Tsueda K (1986) Total airway occlusion and superior vena cava syndrome in a child with an anterior mediastinal tumor. *Anesth Analg* 65: 1079–1082
12. Davis PF, Shevland JE (1985) Superior vena caval obstruction: an analysis of seventy six cases with comments on safety of venography. *Angiology* 36:354–357
13. Davenport D, Ferree C, Blake D et al (1976) Response of the superior vena cava syndrome to radiation therapy. *Cancer* 38:1577–1580
14. Nitschke R, Acker S, Campbell D et al (1975) Superior vena cava syndrome. *JAMA* 233:1354–1355
15. Schindler N, Vogelzang RL (1999) Superior vena cava syndrome. Experience with endovascular stents and surgical therapy. *Surg Clin North Am* 79:683–687