Kartagener's syndrome – anaesthetic considerations for ENT surgery

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Kartagener’s syndrome – anaesthetic considerations for ENT surgery

Zespół Kartagenera – anestezjologiczne uwagi w chirurgii laryngologicznej

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ABSTRACT

Kartagener’s syndrome is a rare autosomal recessive disorder presenting a triad of sinusitis, bronchiectasis and situs inversus with dextrocardia. It occurs in 50% of patients with situs inversus. The most important anesthetic implications of Kartegener’s syndrome surgery are assessment of pulmonary and cardiac structure and function. We present a case of 43-year-old woman with chronic rhinosinusitis with polyps and bilateral secretory otitis media. The chest radiograph and CT scans showed dextrocardia and situs inversus with chronic bronchitis without bronchiectasis. Spirometry showed forced expiratory volume in one second (FEV1) of 2.66 L and forced vital capacity (FVC) of 3.62 L. Electroechography showed no cardiac abnormalities with 55–60% of EF. The anesthetic implications of Kartagener’s syndrome are varied. The regional or general anesthesia might be involved with sinus surgery, ear surgery, pulmonary surgery, infertility or abdominal and cardiac surgery. The main anesthetic considerations among patients with Kartagener’s syndrome are related to the pulmonary function which include preoperative respiratory infections due to bronchiectasis. We should also monitor potentially occluded congenital heart diseases. Kartagener’s syndrome is a rare disease and when the patient need an operation we have to consider surgery with regional or general anesthesia. The general anesthesia would be safe after complete preanaesthetic examination of the patient. The ECG, chest CT scans, spirometry and echocardiography are mandatory before the operation.

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**Introduction**

Kartagener’s syndrome is a variant of the immotile cilia syndrome and occurs in 50% of patients with situs inversus [1]. It is a rare autosomal recessive disorder presenting a triad of sinusitis, bronchiectasis and situs inversus with or without dextrocardia [2, 3]. Chronic ear, sinusitis and chronic respiratory infections as well as male infertility are the most common, but a congenital heart disease could also be observed [3–6].

The proper diagnosis and treatment of pulmonary infection can prevent irreversible damage of the lungs [1, 4, 7]. The most important anaesthetic implications of Kartagener’s syndrome surgery is an assessment of potential respiratory or cardiac complication [1].

Nasal mucosa, paranasal sinuses, middle ear, eustachian tube, pharynx as well as the lower respiratory tract down to the respiratory bronchioles is covered by unciliated epithelium resulting from ineffective mucociliary clearance, undergoing to ENT surgery.

**Case report**

A 43-year-old caucasian female was admitted to our hospital because of chronic rhinosinusitis with polyps and headache. From early childhood she was treated for chronic discharge from nose and had suffered from hearing loss and episodes of respiratory mild infections.

Cardiovascular examination of a patient demonstrates the heart sounds which were heard best on the right side of the chest. Physical examination showed totally obstructed nasal passages with signs of chronic purulent sinusitis and polyps which were well presented at a CT scans (Fig. 1). Ear examination showed signs of secretory otitis media with thickened ear drum and conductive hearing loss at the level of 60 dB for the right side and 70 to 90 dB for the left side. Tympanometry for both ears was typical for secretory otitis media with type B. At the CT scans of the temporal bone, both mastoid process and middle ears cavities were obstructed with pathological mass without damage to the ossicular chain (Fig. 1). The chest roentgenography and CT scans showed dextrocardia and situs inversus (situs inversus totalis) with chronic bronchitis of the lung but without bronchiectasis (Fig. 2). A spirometry showed forced expiratory volume in one second (FEV1) of 2.66 L (91% of predicted value) and forced vital capacity (FVC) of 3.62 L (104% of predicted value). No changes in blood gas analysis were found. ECG demonstrated sinus rhythm and inversion of the P wave in lead I with a deep Q wave and inversion of the T wave. ECG with reversed leads showed no abnormalities. Echocardiography showed no cardiac abnormalities with 55–60% of EF.

The patient was operated twice. First in 2008 a FESS surgery was done because of chronic rhinosinusitis with polyps. The second operation of chronic otitis media was done 9 months after the first one.

Pre-anaesthetic evaluation with all tests, which had been performed, helps to prepare a patient for safe anesthesia and surgery. After intravenous pre-operative antibiotic prophylaxis (cefazolin sodium) induction of general anesthesia was achieved by administration of intravenous anesthetic (propofol, remifentanil) followed by vecuronium in routine dose to facilitate endotracheal intubation. Routine monitoring was established (continuous electrocardiography ECG with mirror image of normal placement of electrodes, pulse oximetry, NIBP monitoring, capnography). Maintenance of anesthesia was achieved with sevoflurane and remifentanil in the target control infusion (TCI) system using an infusion pump Orchestra Base Primea (Fresenius Kabi AG, Germany).

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**Fig. 1 – CT scans of the middle ear and paranasal sinuses showed: soft tissue mass in the middle ear cavity on both sides and totally opacification of the sinuses and nasal cavity with soft mass**
Controlled ventilation with a respiratory rate (RR) of 8 per minute and tidal volume (TV) of 8 ml/kg was adjusted. At the end of surgery, neuromuscular blockade using neostygmine was reversed. The patient was extubated and remained in post-anesthesia care unit (PACU) for 24 hours, where standard care was performed moreover respiratory physiotherapy and exercise to clear bronchial secretions to prevent bronchitis was applied. Her postoperative course was uneventful and she was sent home after a few days. The patient is under pulmonological and laryngological postoperative follow up.

Discussion

Kartagener’s syndrome was first described in 1933 as an association of situs inversus, chronic sinusitis and bronchiectasis [8]. The Kartagener’s syndrome is caused by ciliary dysfunction which result from the defect of ultrastructure of cilia. The absence of the inner or outer dynein arms impair the coordinated ciliary motion and are responsible for mucus retention leading to chronic infections [1, 7, 9, 10].

The anaesthetic implications of Kartagener’s syndrome are varied. Both regional or general anesthesia might be involved [1, 7]. The advantage of regional anaesthesia, especially epidural anesthesia and analgesia appearing to be superior because of the lower risk of high motor block [7].

The main anesthetic considerations among patients with Kartagener’s syndrome are related to the pulmonary function which include preoperative respiratory infections due to bronchiectasis [1, 7, 11]. It is often necessary to delay surgery if respiratory infection is present. This is very important especially in the older patients which presented usually severe bronchiectasis [12]. The chest CT, a spirometry examination and blood gas analysis should be obligatory before the operation.

Bronchiectasis can be categorized as a chronic obstructive pulmonary lung disease (COPD) [13]. Chronic obstructive pulmonary disease (COPD) is considered independent risk factor for mortality and major pulmonary complications after surgery [14]. Bronchoconstriction and hyperdynamic inflation during mechanical ventilation could be prevented by using volatile anesthetics agents, which are known to relax directly bronchial and vascular smooth muscles [15]. The respiratory rate should be low to allow increased expiratory time [14, 16]. Patients with combined bronchospastic and coronary artery diseases might benefit from the administration of alpha-2 adrenergic agonists [14].

Because the lack of effective pain relief as well as drowsiness and respiratory depression associated with the use of long-acting systemic opioids may result in diminished chest expansion and ineffective cough, leading to basal atelectasis, hypoxaemia and nosocomial infection, use of short-acting opioid during general anesthesia and adequate postoperative pain management is very important [1, 5, 7].

When we have dextrocardia with situs inversus the heart is mostly normal. Dextrocardia with complete situs inversus occurs in about 2 per 10 000 births and in that cases congenital heart diseases could occured in less than 3% [1]. If dextrocardia occurs with situs solitus or ambigus which is less common (1 per 20 000 births) the incidence of congenital heart disease is very high, nearly 90% [1, 7]. A very high rate of heart disease is also observed in levocardia with situs inversus [1].

Conclusions

When we plan the surgery for chronic ear or sinusitis we should think about primary ciliary dyskinesia or Kartagener’s syndrome and this pathology have to be recognised in pre-operative period.

Kartagener’s syndrome is a rare disease and when the patient need an operation we have to consider surgery with regional or general anaesthesia. The general anaesthesia would be safe after complete preanaesthetic examination of the patient. The ECG, chest CT scans, spirometry and
echocardiography are mandatory before the operation. The multidisciplinary team working could minimalise the adverse complications.

Authors’ contributions/Wkład autorów

According to order.

Conflict of interest/Konflikt interesu

None declared.

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